

Factor level testing

Clotting factor level tests measure how much factor VIII or IX the body is producing. This will help to understand the risk of abnormal bleeding.

Who should have factor level testing?

Clotting factor level testing is recommended for:

- All girls and women who have the gene alteration for haemophilia
- Girls and women who are very likely to have the gene alteration because of their family's history of haemophilia
- Girls and women who have bleeding symptoms that might suggest low factor levels.⁵

Checking factor levels will help to determine if you or your daughter need a treatment plan.

It is also often the first step in finding out if a female is a haemophilia carrier. Lower than normal factor levels usually indicate a female is a haemophilia carrier if there is already a family history of haemophilia.

However, if you have normal clotting factor levels, you should not take this result to mean you aren't a haemophilia carrier. It just means you are producing normal levels of clotting factor. You will need genetic testing to confirm whether you carry the gene alteration or not.

When to have factor level testing

Ideally factor level testing should be done early in life, in case a girl is likely to have bleeding problems that need to be managed. The exact timing of the blood test is an individual decision but is recommended before she has her first menstrual period (menarche).

Factor level testing is not recommended for an embryo or fetus as it is a complex procedure with risks.

For young children, a good time to collect the blood for a clotting factor level test can be at the same time as a blood test for another reason. If your daughter is already going to have a blood test, you can contact the Haemophilia Treatment Centre who can arrange the form to collect the extra sample of blood for clotting factor testing at the same time.

As an adult you may also have factor level testing at the same time as other blood tests such as genetic testing.



Factor level testing over a lifetime

When symptoms appear

Factor level testing may be recommended if a girl or woman has abnormal bruising and bleeding which suggests haemophilia.

Periods (menstruation)

Factor level testing is important before a girl starts menstruating (getting her period) or if you are having heavy periods.

This can give a girl and her parents the opportunity to be prepared and well-informed about the possibility of heavy periods before her periods start. It also helps with managing heavy periods once they occur. The initial treatments for heavy periods are generally similar whether or not you have a bleeding disorder, but there may be other treatment options that are appropriate if your factor level is low. Managing heavy periods with a treatment plan can make a big difference not only to your health, but also to study, work and generally participating in and enjoying life.

Surgery, medical procedures or dentistry

If you are having planned surgery or medical or dental procedures that pierce the skin, no matter how minor, it is essential that you have your clotting factor levels tested. If your level is low your Haemophilia Treatment Centre will put a management plan in place to make sure the procedure can proceed safely.

Pregnancy

It is particularly important to have had clotting factor level testing **before** you become pregnant, or as soon as possible if you did not plan the pregnancy. This is to manage any potential bleeding throughout pregnancy and childbirth:

- with invasive procedures where there is a risk of bleeding, such as in procedures with IVF (in vitro fertilisation) and prenatal diagnostic testing of the fetus
- preparing for bleeding risks during childbirth and after delivery.

Factor VIII deficiency (haemophilia A): If your factor VIII level is low and you become pregnant, you will also need more factor level testing as your pregnancy progresses. Usually factor VIII levels increase during pregnancy, so another test might need to be done in your third trimester to see if the level is high enough for a safe birth and to prevent bleeding after delivery. If not, replacement factor therapy, tranexamic acid or other treatment might be needed.

Factor IX deficiency (haemophilia B): Factor IX does not change during pregnancy, so testing does not need to be repeated.

Other times?

Talk to your haemophilia team about other times you might need to have your clotting factor level done again. This would depend on your individual situation.

Factor levels and severity

The normal level of factor VIII or IX in a person's blood is between 50% and 150%

Severity and factor level

What to expect if you are female

All severity and factor levels

- Likely to bruise easily and have prolonged bleeding after minor cuts
- Likely to have bleeding problems after a bad injury, tooth extractions, surgery or medical procedures that pierce the skin

Mild haemophilia

5 – 40% of normal clotting factor

- May have heavy menstrual bleeding (heavy periods)
- Might have bleeding problems with childbirth
- Other than periods, might only have bleeding problems requiring medical attention very occasionally

Moderate haemophilia

1 – 5% of normal clotting factor

- Likely to have heavy menstrual bleeding (heavy periods)
- Sometimes have bleeding problems with childbirth
- Might have bleeding problems with minor injuries, such as sporting injuries
- Occasionally have a bleeding episode for no obvious reason ('spontaneous bleeds')

Severe haemophilia

Less than 1% of normal clotting factor

- Likely to have heavy menstrual bleeding (heavy periods)
- Likely to have bleeding problems with childbirth
- Often have bleeding into joints, muscles and soft tissues
- Can have bleeding episodes for no obvious reason ('spontaneous bleeds') as well as after surgery, dental work or injuries including minor bumps and knocks.



Women and girls can have very variable symptoms with haemophilia.

“As long as I can remember, I was told that I was a carrier. My older brother didn't have haemophilia. We wrestled, rode our bikes, fought and were just kids that loved to play outside. And yes, very often I would end up covered in bruises from head to toe!”

“I have haemophilia A with levels of 28%. I do have bleeding symptoms. I have had heavy periods, bruised many times without impact, had post-partum haemorrhages [after childbirth] and prolonged bleeding after cuts and surgeries.”

“I am a female with mild haemophilia A. I was only diagnosed following my son's diagnosis. If I hadn't been tested, I would never have known I was a carrier and had the condition. I get very heavy periods but I didn't think anything of it and once I had very minor surgery with an internal bleed afterwards. They were the only occasions where I have been affected.”

What does the factor level test involve?

“Factor level testing is just like a normal blood test – it's painless and quick. The process isn't scary. You're fully informed by your haematology doctor and nurse about the results and what to do if the test returns positive.”

Factor level testing is a simple blood test.

Factor level testing can be done at the blood collecting service at the same hospital as your Haemophilia Treatment Centre or you may be referred to a local pathology service.

Only a small amount of blood is taken – 3.5mls or around a teaspoon or less.

Results

When you are given your results by the haematologist or the nurse practitioner at the Haemophilia Treatment Centre, they will explain what the results mean. There may be some variation in the results, depending on the laboratory and your Treatment Centre will need to interpret this for you.

Michelle's story

Although Michelle had a family history of haemophilia, it took many years to connect the dots with her bleeding symptoms. But it was a journey that resulted in a great improvement to her quality of life.

'Haemophilia B is in my family. We often spoke at family gatherings about my grandfather who was diagnosed in the 1920s at around age 12 after falling out of a tree and spent many months in hospital. He was told by a doctor to "come back and find me if you live to see 21" – which he did.'

'I was studying Science at university, majoring in genetics, when I read in a textbook that haemophilia B is X-linked and as my grandfather and cousin have haemophilia B, there was a chance I carried the gene.'

Over the years Michelle saw a few GPs about her heavy menstrual bleeding and anaemia and raised her family history of haemophilia. 'The GPs considered haemophilia too rare and unlikely to affect women to agree to a factor level test, let alone a genetic test.' Eventually in her 30s she sought help from a women's health clinic and was referred to the specialist Haemophilia Treatment Centre.

'I had counselling from the Haemophilia Treatment Centre clinical nurse consultant prior to having the genetic test.'

'The clinical nurse consultant was very approachable, sensitive, knowledgeable and professional. She immediately put me at ease.'

After her diagnosis, Michelle had a hormone-releasing IUD implanted and no longer has bleeding symptoms.



Treatment plan

Each person affected by haemophilia will have their own treatment plan worked out with their haematologist (specialist doctor qualified to manage blood diseases) or haemophilia nurse practitioner that takes into account their individual type of haemophilia and bleeding pattern along with the best option for each situation. Treatment for women and girls may also involve a gynaecologist (specialist women's health doctor) if heavy menstrual bleeding is a problem.

Treatment can be given to:

Prevent bleeding episodes, for example:

- regular 'prophylaxis' (preventive) treatment
- or treatment before surgery, medical or dental procedures
- or treatment before and after childbirth
- Some women find hormonal treatments such as contraceptive pills, implants or IUDs helpful in preventing bleeding.

Control a bleeding episode **once it has started**
This is called '**on demand**' therapy.

Types of haemophilia treatment can include factor replacement therapy, non-factor therapies, hormonal treatments, and antifibrinolytic drugs such as tranexamic acid. You may be prescribed different types of treatment at different times.

Researchers are always looking for new ways to treat haemophilia and new treatments are now in development and becoming available. This includes clinical trials for gene therapy, which may in the future be able to cure haemophilia.

Australian Bleeding Disorders Registry (ABDR)

If you have bleeding symptoms, registering and staying in contact with your local Haemophilia Treatment Centre (HTC) is an important part of managing your bleeding disorder, even if you receive most of your care from another doctor, such as your general practitioner (GP).

If you are diagnosed with haemophilia or as carrying the gene, you may be asked by your HTC to register in the **Australian Bleeding Disorders Registry (ABDR)**. This is the online system used by HTCs nationally to manage and record the treatment and care of their patients.

The ABDR is an important resource for people with bleeding disorders.

- Your diagnosis and treatment data is centralised and easily accessible by your treating team
- With your permission, your full data can be shared with another HTC if you move from a children's to an adult HTC, move interstate, or are travelling within Australia
- Statistics from the ABDR can support research into understanding and treating bleeding disorders, along with planning for treatment product supply nationally.

Ask your HTC about an **ABDR patient card**.

This is a wallet card which explains:

- Your diagnosis and severity
- Your treatment plan
- Who to contact for further medical advice.

Keep your ABDR patient card on you for quick reference and to show to other health professionals treating you.



Haemophilia Treatment Centre
City Hospital, OZ CITY, OZ
Haemophilia Centre Ph: 01 2111 3111
On-Call Haematologist (A/H) Ph: 01 2111 3000

Jane Citizen

DOB: 1/1/2001 MRN: 0100101 ABDR ID: 100200

Diagnosis: **Factor VIII Deficiency (Haemophilia A)**
Severity: Mild Factor Level: 12%

Card Issue Date: 16/03/2021

Treatment Guidelines

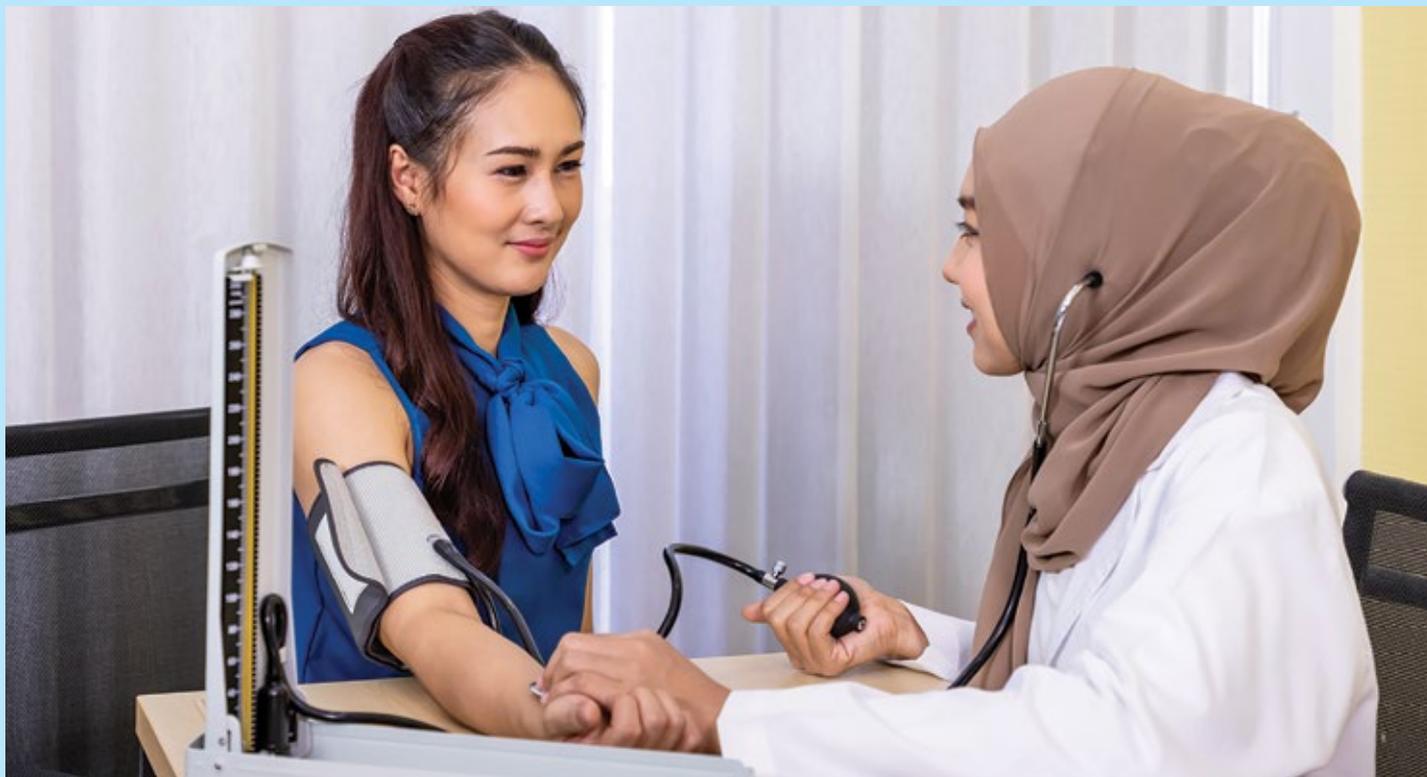
Product:
RECOMILISTATE (Extended Half-Life Recombinant Factor VIII)

Treatment Comments:
Treat **PROMPTLY** with Recombinant factor VIII (8) to avoid serious complications. Consult with Centre or Haematologist on call for dosing advice, and follow-up treatment.
Will require cover for trauma, medical or dental procedures or surgery.

Factor doses rounded up to nearest vial size.

MyABDR: www.blood.gov.au/myABDR MyABDR Support Ph: 13 000 25663

Another important tool is the **MyABDR** app and website. If you are using haemophilia treatment products at home, you can use MyABDR to record your treatments and bleeding episodes and share the information with your HTC. Your MyABDR entries will be added directly to your record in the ABDR.



What can you tell your other doctors?

Bleeding disorders like haemophilia are relatively rare conditions. Most doctors and dentists may not have treated female patients with bleeding disorders before and will not be familiar with current treatment guidelines for haemophilia.

- Give your other doctors, dentist and health care providers a copy of any letters explaining your diagnosis and the contact details of your Haemophilia Treatment Centre. Ask them to liaise with your HTC.
- Show them your ABDR patient card, if you have one.
- Always inform your doctor, dentist or surgeon you have a bleeding disorder before having any medical, dental or surgical procedures, no matter how minor.
- Before you have any procedures, contact your HTC and discuss the medical support you may need to prevent bleeding complications.
- Your HTC might also have specific brochures you can take with you, eg on surgery or dentistry.
- Before you start taking anything prescribed by your doctor, naturopath or other health practitioner, check with them whether it is safe for someone with a bleeding disorder.

Where can your other doctors get information?

Your other doctors may wish to look at current evidence-based clinical care guidelines. In Australia these are produced by the Australian Haemophilia Centre Directors' Organisation (AHCDO) and can be downloaded from their website – www.ahcdo.org.au.

With your permission, your treating haematologist (specialist bleeding disorders doctor) or the team at your HTC would be happy to speak with your doctors and other health professionals about your diagnosis and treatment plan and liaise with them about your care.

Sharri's story



You often hear about mothers of children with haemophilia devoting huge amounts of time to getting their child's medical issues sorted out. Frequent appointments at haematology, physiotherapy, pathology and elsewhere take time in an already busy world for parents. This is one of the reasons Sharri, whose son has haemophilia, found it hard to get on top of her own medical situation – which turned out to be a little complicated.

'After he was diagnosed at a few days old and the dust settled (several years later) I took myself off to get blood levels checked for the very first time. I didn't know I was a carrier before I had my son. My factor levels were 38% which I was told was not great but if I required any surgery probably best to check in with them first. As I was fit and healthy, I was fairly sure any surgery was not in my plans for the next 20 or so years, so I just carried on not thinking about it much.'

'I continued my haematology checks and the last one had my factor levels at just 17% and I was also diagnosed as having mild von Willebrand disorder. I still wasn't too worried as it didn't really affect me day to day. I had a hormone-releasing IUD implanted, which managed the curse of my menstruation.'

Things took a bit of a twist when Sharri needed surgery.

'I told my surgeon about my bleeding issues and his clinic communicated with the Haemophilia Treatment Centre. Turns out I definitely needed treatment product and a pre- and post-surgery treatment plan was written up. I required factor replacement therapy over the five days I was in hospital and could not go home until the last blood test said my levels were good and everyone was happy.'

Sharri hopes others will learn from her experience by taking the time to look after themselves.

'I want to encourage women who are carriers to get their factor levels checked. It has been an interesting learning experience for me because for so long it has been all about my son and managing his care, but this time - for the first time - it has been all about me.'

Sources

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NB: All photos in this booklet are stock images.

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More information

To find more information about haemophilia and carrying the gene alteration, or to find out how to get in touch with your local Haemophilia Foundation or a specialist Haemophilia Treatment Centre, contact:

Haemophilia Foundation Australia

7 Dene Ave Malvern East Victoria 3145

T: 03 9885 7800 Toll free: 1800 807 173

E: hfaust@haemophilia.org.au

Or visit the HFA website: www.haemophilia.org.au

Important note

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