

MY LIFE WITH **HEMLIBRA[®]** (emicizumab)

A guide for people with severe congenital haemophilia A (FVIII <1%) or moderate congenital haemophilia A (FVIII \geq 1% and \leq 5%) with severe bleeding phenotype, who have been prescribed Hemlibra[®] (emicizumab)



My Hemlibra: My guide

You have been prescribed Hemlibra to help manage your haemophilia A.

Inside this booklet, you'll find information to help you get the most from your Hemlibra treatment.

In addition to this booklet, before administering Hemlibra you should read the package leaflet contained in the box with your medicine, the guide for patients/carers and the patient alert card your doctor or nurse will have given to you.

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Getting to know Hemlibra

As you begin a treatment, change can feel both exciting and a little daunting. It may take a while for you to get used to your routine with Hemlibra. This booklet is designed to help you understand your condition and how Hemlibra can help you manage it.

Hemlibra is different to factor VIII replacement therapy. It is used for people with severe haemophilia A (FVIII<1%), moderate haemophilia A with a severe bleeding phenotype (FVIII>1- ≤5%) and also for people with FVIII inhibitors. You have been prescribed Hemlibra because you and your healthcare team decided it was right for you; it does NOT mean you have inhibitors to factor VIII.



Hemlibra is not a cure for haemophilia A. It is a prophylactic treatment used to prevent or reduce the frequency of bleeds. In studies to test Hemlibra, some people had no bleeds that needed treatment.^{1, 2}

This booklet may not answer all of the questions you have about Hemlibra. Please ask your doctor or nurse if there is anything else you would like to find out.

Haemophilia A

Haemophilia A is a bleeding disorder that means your blood cannot clot properly.

Blood contains lots of clotting factors, which interact with each other in a sequence to make a clot form. This is called the 'clotting pathway'.

People with haemophilia A do not have enough of one of these clotting factors – factor VIII.

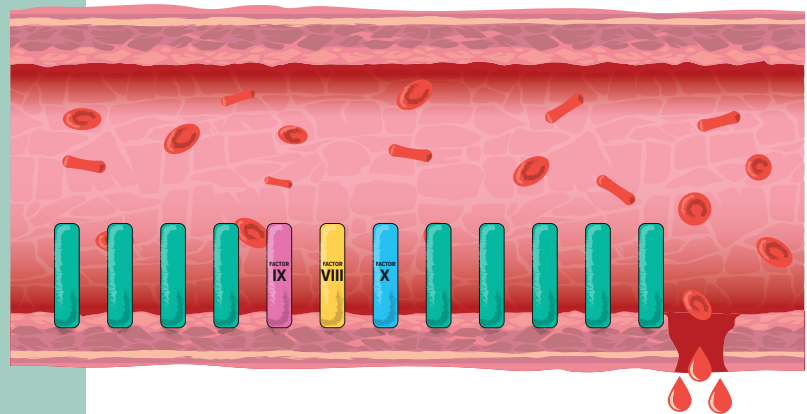
Haemophilia A can be described as mild, moderate or severe, depending on how much FVIII is in the blood.

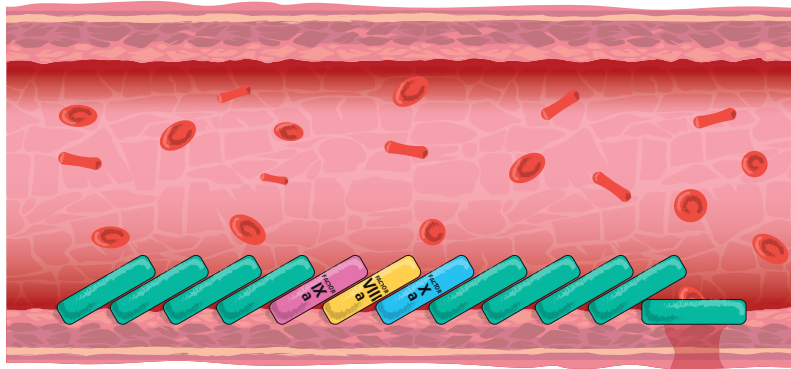
People with severe haemophilia A have <1% FVIII and people with moderate haemophilia A have between 1-5%.

Some people with moderate haemophilia A can bleed more than others. This is often referred to as having **“a severe bleeding phenotype”**.

The clotting pathway is a bit like dominoes falling

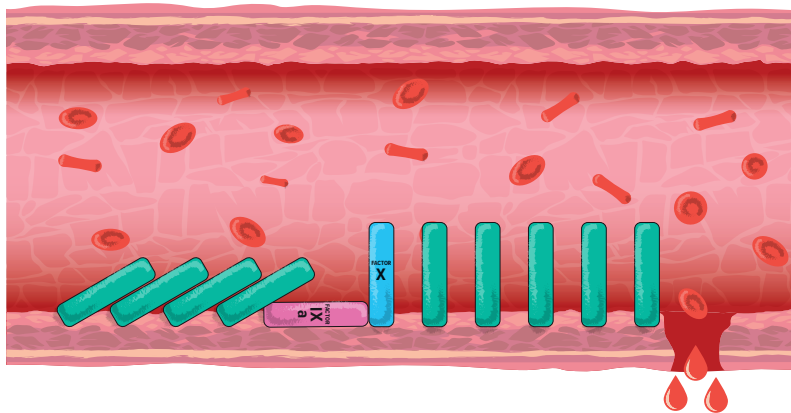
This illustration represents factors in the clotting pathway that are relevant to haemophilia A.





Factor VIII is the link between activated factor nine (factor IXa) and factor ten (factor X).

When factor IXa falls, factor VIII needs to be in place so that it can fall on to the factor X domino.



If factor VIII is missing, the rest of the dominoes will not fall and a clot will not form.

Haemophilia A can be managed with factor VIII replacement therapy.

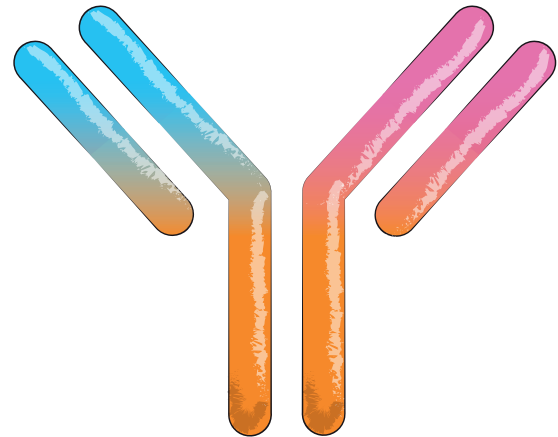
Factor VIII therapy can be used to help blood clot in response to injury. It can also be administered regularly, to help prevent bleeds from happening. Taking a treatment regularly to try and prevent bleeding is called prophylaxis.

Understanding how Hemlibra works

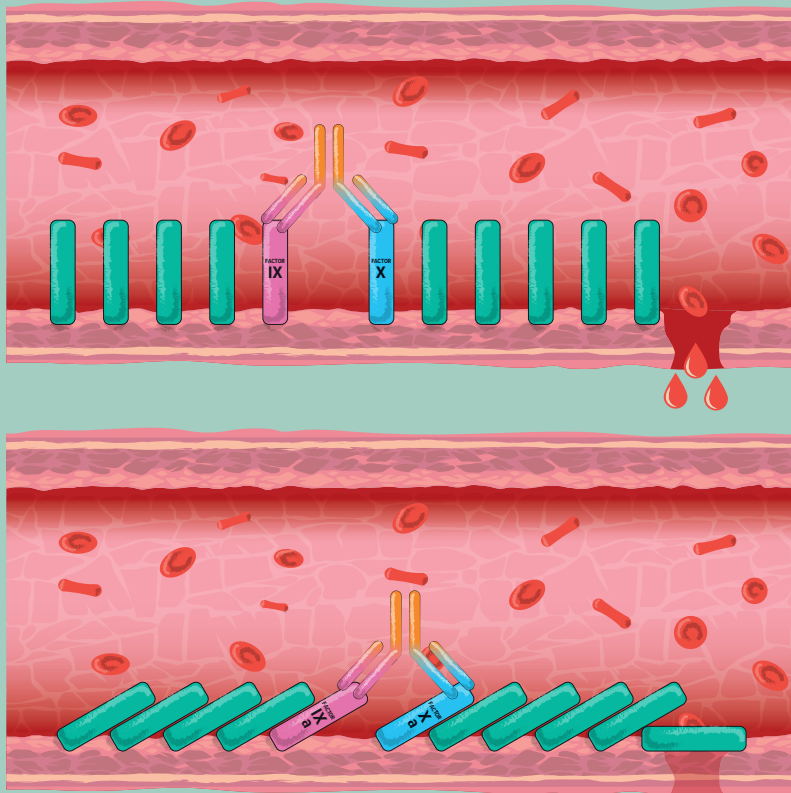
Antibodies are proteins made by the body that stick to substances (antigens). Antibodies can be important in fighting infection. However, they can also be designed to bind to specific substances to manage diseases; these are called monoclonal antibodies.

Hemlibra is a monoclonal antibody designed to help prevent bleeding in people with severe haemophilia A and moderate haemophilia A with a severe bleeding phenotype.

- Hemlibra is different to factor VIII replacement therapy
- Hemlibra is a bispecific antibody, meaning it can bind to two different substances
- Hemlibra binds to factor IXa and factor X. This forms a link so the clotting pathway can continue



**Diagram of the Hemlibra
monoclonal antibody**



Remember the dominoes?

Hemlibra binds to factor IXa and factor X, bridging the gap so the clotting pathway can continue and a clot can form.

How to take Hemlibra

Hemlibra must be taken regularly as prophylaxis



Hemlibra is taken once-weekly for the first 4 weeks (this is called your loading dose) and then once every one, two or four weeks thereafter.



If you have a bleed you should not take additional Hemlibra. See page 20 for more information on managing a bleed.



Your Hemlibra dose is not routinely adjusted according to your blood results. You must not adjust the dose yourself, for example if you are planning on playing any sports.



If you are already using factor VIII prophylaxis this can be continued for the first 7 days of your Hemlibra prophylaxis.

Your dose is based on your body weight

LOADING DOSE

For the first 4 weeks, you take a loading dose of Hemlibra.



ONGOING DOSE

From week 5, you carry on with a regular ongoing dose



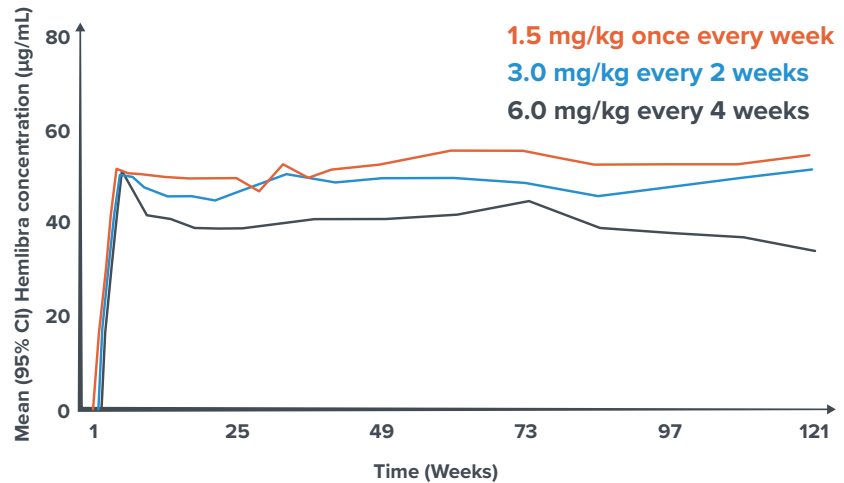
How to take Hemlibra

Hemlibra

Taking Hemlibra regularly maintains a consistent level in your blood.

After the initial loading dose, blood levels of Hemlibra remain stable between doses, whether you are taking your ongoing dose once every week, every 2 weeks, or every 4 weeks.

This means that you do not need to take additional doses of Hemlibra to cover any planned physical activity.



(adapted from Callaghan et al, blood 2021)

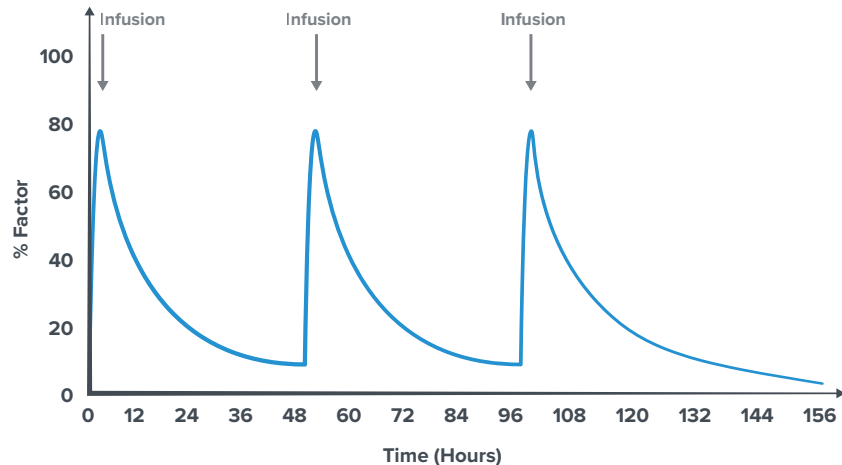
Factor VIII replacement therapy

You may have previously taken factor VIII replacement therapy as prophylaxis for your severe or moderate haemophilia A, or know someone who does.

The level of factor VIII replacement therapy rises quickly after infusion and then decreases, as shown by the peaks and troughs on this graph.

Representation of factor VIII levels after IV administration

(note; This graph illustrates the differences in peaks and troughs of FVIII replacement therapies. It does not represent any specific products and is not representative of all FVIII replacement products.)



(adapted from Castaman G, Linari S. *Exp Opin Drug Metab Toxicol.* 2018;14:143-151)

How to take Hemlibra

Hemlibra is given as an injection under the skin, called a **subcutaneous injection**. This may be different from what you're used to.

- **Your haemophilia team will teach you how to perform subcutaneous injections**
- **Your haemophilia team will calculate your dose of Hemlibra based on your weight**
 - Let them know if you lose or gain weight so that they can adjust your Hemlibra dose
 - Do not try to calculate your dose of Hemlibra yourself
- **The acronym **PARIS** can help you to remember each step correctly and make injecting Hemlibra more comfortable**

AS A REMINDER, there are detailed instructions in the Hemlibra package leaflet that comes with your medication. This includes how to prepare your vial(s) and how to inject Hemlibra. **A reminder video is available online at www.takingemicizumab.com. You can also access this from the QR code printed on the last page of this booklet.**

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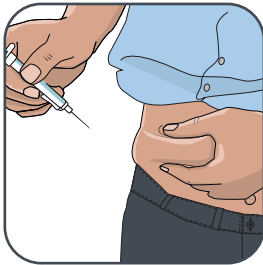
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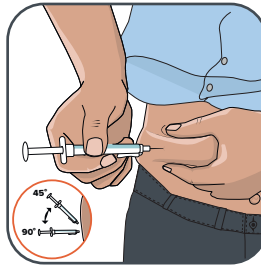
S

PINCH



Pinch the skin at the selected injection site.

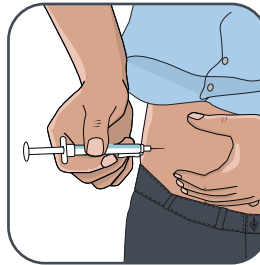
ANGLE



Angle the needle at 45° to 90° and insert with a quick, firm action.

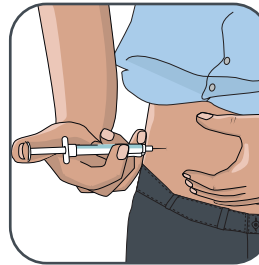
Do not hold or push on the plunger while inserting the needle.

RELEASE



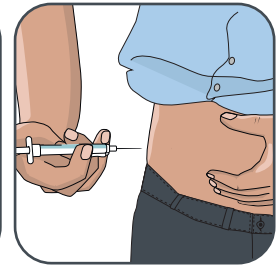
Release the pressure between the pinched skin whilst holding the syringe still, in the same position.

INJECT



Inject all the prescribed dose slowly by gently pushing the plunger all the way down.

SLIDE



Slide the needle gently out from the injection site at the same angle as inserted.

Preparing for Hemlibra injections

- Wash your hands well with soap and water before preparing and administering Hemlibra
- Before use, allow the vial(s) to reach room temperature for about 15 minutes on a clean flat surface away from direct sunlight
- For each injection, use a different area of the body to the one you used last time. You should use a different injection site for each injection, at least 2.5cm away from the area you used for your previous injection. This also applies if you need to have more than one injection for your prescribed dose
- Clean the chosen injection site area using an alcohol wipe and let the skin dry for at least 10 seconds
- Do not inject into areas that could be irritated by a belt or waistband. Do not inject into moles, scars, bruises, or areas where the skin is tender, red, hard or the skin is broken

Tips for remembering your injections



Set reminders on your phone or computer.



Schedule your injection day around a regular activity.



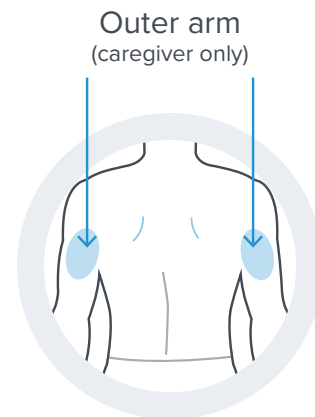
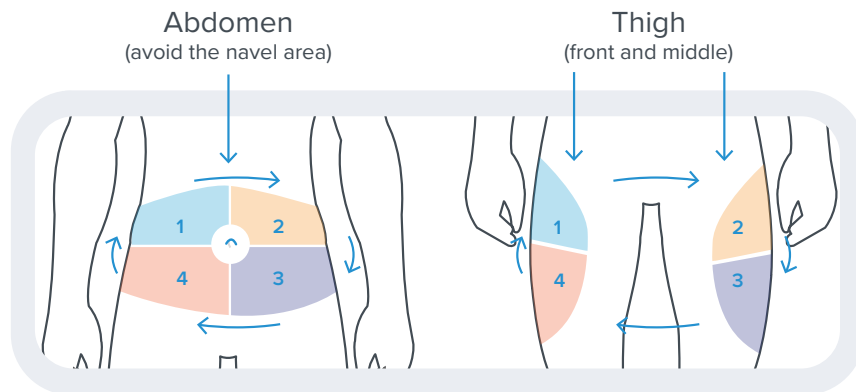
Continue to use Haemtrack® to keep a record of each time you use Hemlibra.

Where to inject Hemlibra

The recommended injection sites are the abdomen and thigh. Additionally, the outer arm, when administered by a caregiver.

These sites are recommended because they have an easily accessible area of fatty tissue just below the skin.

Do not inject into areas that could be irritated by a belt or waistband. Do not inject into moles, scars, bruises, or areas where the skin is tender, red, hard or if the skin is broken



Getting started

Hemlibra is always prescribed as routine prophylaxis. It is essential to take Hemlibra regularly to have a better chance of preventing bleeds.

DOs



Take Hemlibra regularly as instructed



Continue to take Hemlibra, even if you haven't had a bleed in a while



If your prescribed dose is more than 2 mL you will need to have more than one subcutaneous injection of Hemlibra. These injections must be given on the same day with each injection at least 2.5 cm away from the area used for the previous one

DON'Ts



Do not take extra Hemlibra, even if you do have a bleed



Do not adjust your dose of Hemlibra yourself

Continuing Hemlibra

Once you have taken Hemlibra for 4 weeks at the loading dose you will continue to inject Hemlibra regularly, as instructed by your haemophilia team.

Regular blood tests to check your factor levels are not needed when you are taking Hemlibra. Your dose is not based on your clotting factor levels.

If you do need to have any laboratory or blood tests, tell your doctor you are taking Hemlibra, as it can interfere with some laboratory tests, leading to inaccurate results.



Hemlibra clinical trials



*Hemlibra has been tested in clinical trials
in people with severe and moderate haemophilia A .*

*Hemlibra treatment
was tested in people who
previously used either:*



*On-demand
treatment with
factor VIII therapy*

OR



*Prophylaxis
with factor VIII
therapy*

*Findings from these
clinical trials suggest
that taking Hemlibra
can help prevent
bleeding or reduce the
number of bleeding
episodes*

Mahlangu J, et al. *N Engl J Med*. 2018;379:811–822
Negrier C, et al. *Lancet Haematol* 2023. Mar;10(3)e168-e177

Possible side effects

Like all medicines, Hemlibra can cause side effects, although not everyone gets them.

● Very common: may affect more than 1 in 10 people

- A reaction in the area where the injection is given (redness, itching, pain)
- Headache
- Joint pain

● Common: may affect up to 1 in 10 people

- Fever
- Muscle aches
- Diarrhoea
- Itchy rash or hives (urticaria)
- Skin rash

● Uncommon: may affect up to 1 in 100 people

- Destruction of red blood cells (thrombotic microangiopathy)
- Blood clot in a vein behind your eye (cavernous sinus thrombosis)
- Severe damage of the skin tissue (skin necrosis)
- Blood clot in a vein near the surface of the skin (superficial thrombophlebitis)
- Swollen face, tongue and/or throat and/or difficulty in swallowing, or hives, together with difficulty in breathing which are suggestive of an angioedema
- Lack of effect or decreased response to treatment
- Allergic reaction

Other important information about Hemlibra

Antibody formation (immunogenicity)

You may notice that bleeding is not being controlled with your prescribed dose of Hemlibra. This could be due to the development of antibodies to this medicine.

Talk to your doctor immediately if you notice an increase in bleeds. Your doctor may decide to change your treatment if this medicine stops working for you.

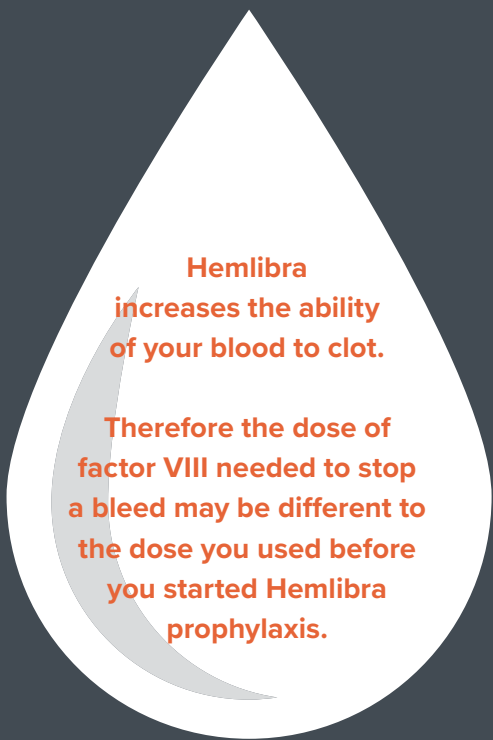
If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the package leaflet. Please report suspected side effects to the MHRA through the Yellow Card scheme, via the Yellow Card website www.mhra.gov.uk/yellowcard, or the free Yellow Card app available in the Apple App Store or Google Play Store. You should also report side effects to Roche Products Ltd. by emailing the Roche Drug Safety Centre at welwyn.uk_dsc@roche.com or calling +44 (0) 1707 367554. By reporting side effects, you can help provide more information on the safety of this medicine.

Being prepared to manage a bleed

IF YOU HAVE A BLEED, DO NOT TAKE EXTRA HEMLIBRA

- It is very important you know how to manage a bleed while you are taking Hemlibra
- If your bleed requires treatment, you will need to use factor VIII replacement therapy. Your haemophilia team will explain when to treat a bleed and the correct dose of factor VIII replacement therapy to use
- If you are using HealthID or have a MedicAlert Bracelet or necklace, you could consider updating your information

The package leaflet contained in the box with your medicine, the guide for patients/carers and patient alert card also have guidance for managing a bleed while you are taking Hemlibra



**Hemlibra
increases the ability
of your blood to clot.**

**Therefore the dose of
factor VIII needed to stop
a bleed may be different to
the dose you used before
you started Hemlibra
prophylaxis.**

What to do if you have a bleed while taking Hemlibra

Please keep this booklet in a safe place so you can refer back to your emergency plan. There is space to write down your contacts and for your doctor or nurse to write down a reminder of your personalised plan for what to do if you have a bleed.

Emergency contacts:

In an emergency:

Remember to keep this updated, if anything changes.

Signed:

Frequently asked questions

Injecting Hemlibra

Who should I tell if I'm having trouble with subcutaneous injections?

Once you have been trained you should be able to inject Hemlibra at home by yourself or with the help of a caregiver. Tell your doctor or nurse if you are not confident with performing subcutaneous injections.

What if I break a vial or it's not sealed well?

Inspect each vial before use. Do not use if they appear damaged, the cap covering the stopper is missing or if they have been dropped. Contact your haemophilia centre and provide the batch details of the affected vial. Keep the vial in the correct storage conditions as you may need to return it when you next attend your centre.



What if the liquid looks strange?

Before using Hemlibra, check the solution for particles or discolouration. The solution should be colourless to slightly yellow. Do not use if it is cloudy, hazy, discoloured or contains visible particles. Contact your haemophilia centre and provide the batch details of the affected

vial. Keep the vial in the correct storage conditions as you may need to return it to your centre.

I'm having trouble drawing up the liquid (getting it into the syringe).

Hemlibra is a viscous liquid, which means it is quite thick. Initially, you might find it difficult to draw it up into the syringe, until you get used to it. Talk to your doctor or nurse if you continue to struggle.

Do I have to wait to have the injection after drawing up the solution in to the syringe?

No. Once the syringe has been filled with Hemlibra, the injection must be given immediately.

Do not refrigerate the solution in the syringe.

How should I dispose of my sharps, needles, and used vials?

Put any used vials, needles, vial/injection needle caps and used syringes in a sharps bin.

Can I save a part-used Hemlibra vial for my next injection?

No. Only use the vial once. After you inject your dose, throw away any unused Hemlibra left in the vial.

I keep getting soreness after injections, what should I do?

Injection site reactions are a very common side effect that some people get with Hemlibra. Make sure that you rotate the injection site with each injection. If you have signs of redness, itching or pain at the injection site, tell your doctor or nurse.



Frequently asked questions

Taking Hemlibra

Should I tell people I have started taking Hemlibra/changed medications?

Your regular haemophilia A care team and your GP will be updated when you change medications. If you have a MedicAlert bracelet or necklace, or a Health ID profile, you could consider updating your information. It is up to you whether you tell your friends and family about your treatment, but it may help to talk to people that are close to you so that they can support you. It's a good idea to carry information about your medicines, such as your patient alert card, in case you see a different doctor, or need to see a doctor when you are away from home.

Can I get blood-borne diseases from Hemlibra?

Hemlibra is a therapeutic antibody made in the laboratory. It is not made from other people's blood or plasma, so there is very little risk of developing a blood-borne disease.

Are there any other medications that I should not take with Hemlibra, or need to be careful with?

There have not been any studies to specifically test how Hemlibra interacts with other medications. If you were previously taking factor VIII replacement therapy as prophylaxis, this can be continued for the first 7 days of Hemlibra treatment.

Bypassing agents are medicines that help the blood to clot but work differently to factor VIII. They are used to manage haemophilia A if people have inhibitors to factor VIII. Examples of bypassing agents include activated prothrombin complex concentrate (aPCC) and recombinant activated factor VII (rFVIIa). Bypassing agents must be stopped the day before starting Hemlibra treatment.

Clinical experience indicates Hemlibra interacts with aPCC. Serious and potentially life-threatening side effects can occur when aPCC is used in people who are also receiving Hemlibra. Use of aPCC should be avoided unless no other treatment can be used. See the package leaflet contained in the box with your medicine, the guide for patients/carers and patient alert card for additional information.

Can I take more Hemlibra if I wake up with pain?

No. If you think that you are having a bleed, you must not take extra Hemlibra. You may need to talk to your doctor or nurse if the pain is severe.

Should I take more Hemlibra if I am going to play sport?

No, Hemlibra is not an on-demand treatment. Prophylaxis with Hemlibra maintains a consistent level in your blood between doses for the whole time that you are on treatment.

After I have started Hemlibra, should I take factor VIII if I am going to play sport or planning other physical activity?

No. When taken regularly, Hemlibra levels are stable, without any peaks and troughs. This means you should not 'top up' your treatment to cover any physical activity unless you have been specifically told to by your doctor or nurse.

Can I stop taking Hemlibra if I haven't had a bleed for a while?

Great news that you haven't had a bleed in a while! However, you still need to keep taking Hemlibra so that you are protected from future bleeding episodes. Do not stop taking Hemlibra without talking to your doctor or nurse.

I want to start a family, is this OK while taking Hemlibra?

Ask your doctor or nurse for advice before using this medicine if you are trying to conceive. There are no clinical studies of Hemlibra use in people trying to conceive, or of Hemlibra in people who are pregnant.

Can I combine Hemlibra doses?

Different Hemlibra concentrations (30 mg/mL and 150 mg/mL) should not be combined in a single injection when making up the total volume to be injected.

What should I do if I forget to take Hemlibra?

If you forget your scheduled injection, inject the forgotten dose as soon as possible up to the day before the day of the next scheduled dose. Then, inject Hemlibra as scheduled. Do not inject two doses on the same day to make up for a forgotten dose. If you are not sure what to do, ask your doctor or nurse.

How long will I be on Hemlibra for?

Hemlibra is intended for long-term treatment. Usually you will continue taking Hemlibra as long as you and your haemophilia team are happy with how your treatment is going. Some people have been on Hemlibra for over seven years.

Can I drive or use machines while taking Hemlibra?

Hemlibra is not likely to affect the ability to drive or use machines.

Frequently asked questions

Storing your Hemlibra

How do I store Hemlibra at home?

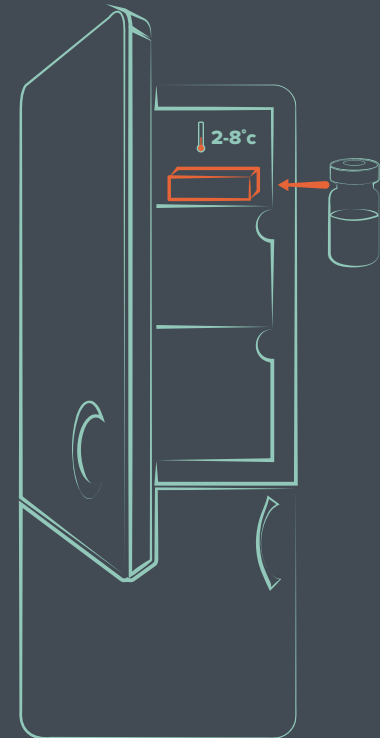
Store Hemlibra in your fridge between 2°C and 8°C in the original box to protect from light. Do not freeze Hemlibra and do not shake the vials. Keep out of the sight and reach of children. Do not use after the expiry date stated on the side of the box and the vial label (the date refers to the last day of the month). Speak to your haemophilia centre if you would like more information on storing medications at home.

Can I take Hemlibra out of the fridge?

Once removed from the fridge, unopened vials can be kept at room temperature (below 30°C) for a total of 7 days. Discard any vial that has been kept at room temperature for more than 7 days or has been in temperatures above 30°C.

Where should I keep my injection supplies?

Keep injection equipment safe and stored away in a cool, dry environment. It is a good idea to keep everything together in a box. Make sure that all supplies are kept out of the sight and reach of children.



Lifestyle

Should I change my diet?

You do not need to change your diet when you are taking Hemlibra, but it is important for anyone with severe and moderate haemophilia A to try to be as healthy as possible; eat well and keep hydrated. The Haemophilia Society and the World Federation of Hemophilia have lots of resources on trying to maintain a healthy diet.

Can I drink alcohol?

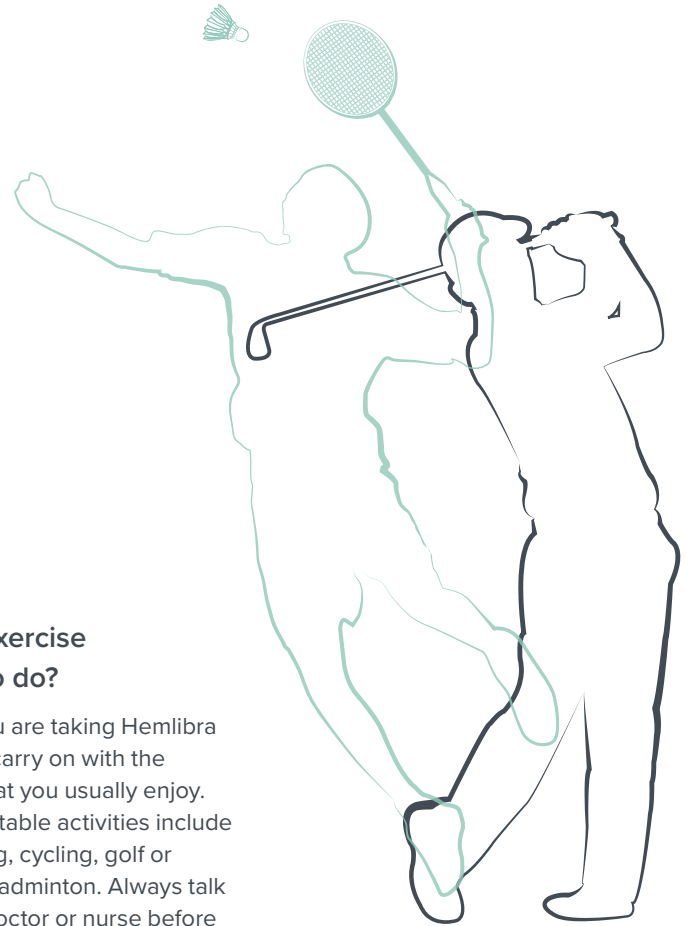
Hemlibra is not affected by drinking alcohol, however it is recommended you stick to government alcohol limits and unit guidelines.

Is smoking OK?

The effects of smoking when taking Hemlibra have not been tested. However, it is best to stay healthy when you have haemophilia A. If you are a smoker, you could consider speaking to your doctor or nurse about ways to reduce or stop smoking.

What exercise is OK to do?

While you are taking Hemlibra you can carry on with the sports that you usually enjoy. Some suitable activities include swimming, cycling, golf or playing badminton. Always talk to your doctor or nurse before starting new activities.¹



1. Srivastava A, et al. *Haemophilia*. 2013;19:e1–47

Frequently asked questions

Travelling with Hemlibra

What do I need to know when travelling with Hemlibra?

If you need to travel with Hemlibra, remember that it can remain out of the fridge for up to 7 days if necessary. However, it is recommended that you store your Hemlibra vials in a fridge whenever possible, even when travelling. Remember to also take your injection supplies and sharps bin with you to safely dispose of your used syringes, needles and vials.

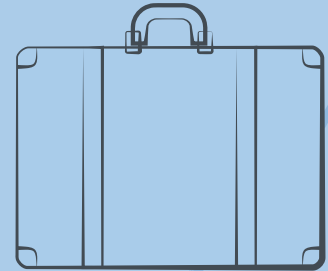
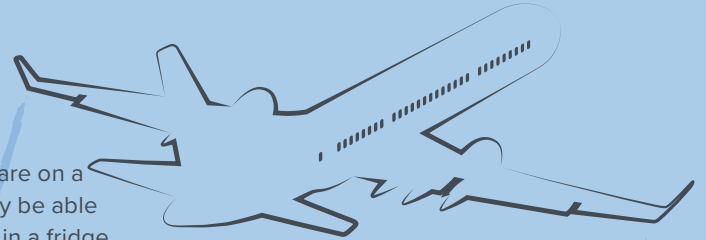
Can I take Hemlibra on an aeroplane?

If you are travelling by aeroplane your doctor or nurse may provide a letter to

give to the airline. If you are on a long flight, the airline may be able to store your medication in a fridge. You should carry your Hemlibra vials with you in your hand luggage so they are not damaged during the flight.

What about my factor VIII?

You may need to carry supplies of factor VIII in case you have a bleed. You will need to carry this in your hand luggage as well. Check the product information provided with the factor VIII replacement therapy and/or talk to your nurse or doctor.



REMEMBER, you should also carry your factor VIII to manage a bleed, in case you have one.

If you have any other questions about Hemlibra, you can write them down here as a reminder to ask haemophilia team at your next appointment.

Glossary

Here are the meanings of some medical terms for you to learn, or to refresh your memory:

Hemlibra is a treatment for haemophilia A that is different to factor VIII replacement therapy. It is used as prophylaxis for people with severe and moderate haemophilia A, and also for people with factor VIII inhibitors. You have been prescribed Hemlibra because you and your healthcare team decided it was right for you; it does NOT mean you have inhibitors.

Antibody

Proteins produced by the immune system that bind to unwanted substances to help the body to remove them.

Clotting

The process by which blood changes from a liquid to a solid, forming a blood clot.

Clotting pathway

The sequence in which clotting factors interact with each other in order to form a clot.

Factor

There are a number of essential blood clotting proteins in the blood known as clotting factors.

Factor VIII

The essential blood clotting protein that is reduced or missing in people with haemophilia A.

Factor VIII replacement therapy

A treatment that replaces the low or missing factor VIII in people with haemophilia A. Factor VIII replacement therapy can be used to help manage a bleed, and it can also be taken regularly as prophylaxis to help prevent bleeds. People who take Hemlibra may need to use factor VIII therapy to manage a bleed.

Haemophilia A

A bleeding disorder where factor VIII is either partly or completely missing. Haemophilia A can be classed as mild, moderate or severe depending on the amount of factor VIII present in the blood.

Immune system

The cells and tissues in the body that provide protection from infections and foreign substances.

Inhibitor

A type of antibody that prevents factor VIII from working.

Monoclonal antibody

Therapeutic antibodies, often referred to as monoclonal antibodies, are designed to bind to specific molecules to help treat a disease or condition.

On-demand

The use of a treatment as needed. In haemophilia A factor VIII replacement therapy is used to manage a bleed. Patients taking Hemlibra may also need to use factor VIII to manage breakthrough bleeds.

For a reminder video on how to administer Hemlibra, access www.takingemicizumab.com using the QR code here



Prophylaxis

The regular use of a treatment to prevent illness. In haemophilia A, prophylaxis is intended to prevent bleeding from occurring or reduce the number of bleeds.

Severe bleeding phenotype

Some people with moderate haemophilia A can bleed more than expected. This is described as having a severe bleeding phenotype.

Subcutaneous injection

A method to deliver medicine into the body that involves inserting a needle into the tissue just beneath the skin.

Special thanks go to...

JULIA SPIRES,

*Clinical Nurse Specialist,
Haemophilia Comprehensive Care
Centre, Great Ormond Street Hospital*

LARA OYESIKU,

*Southern Haemophilia Network
Clinical Nurse Manager, Hampshire
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For their help and guidance during the development of this booklet

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the package leaflet. Please report suspected side effects to the MHRA through the Yellow Card scheme, via the Yellow Card website www.mhra.gov.uk/yellowcard, or the free Yellow Card app available in the Apple App Store or Google Play Store. You should also report side effects to Roche Products Ltd. by emailing the Roche Drug Safety Centre at welwyn.uk_dsc@roche.com or calling +44 (0) 1707 367554. By reporting side effects, you can help provide more information on the safety of this medicine.