



Emicizumab (for people with an inhibitor)

This factsheet is about a treatment called emicizumab or Hemlibra, which is for people with haemophilia A who have an inhibitor to FVIII. Emicizumab has been designed to mimic the way that factor VIII works in the body.

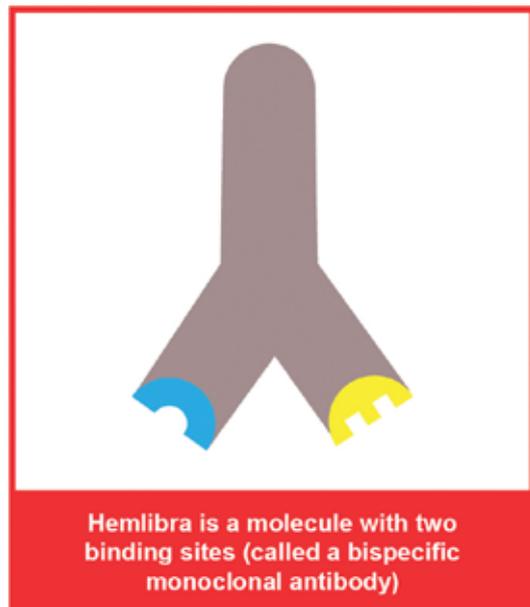
Emicizumab (Hemlibra)

- A new treatment is available for people with haemophilia A who have a Factor VIII inhibitor. It can be used as prophylaxis to prevent bleeds.
- Emicizumab is not a treatment for bleeds, injury or major surgery.
- You have emicizumab as an injection just under the skin either weekly, fortnightly or every 4 weeks. Most people however inject weekly or fortnightly.
- There is no viral infection risk as the product is entirely made in a laboratory.
- There have been some serious side effects reported and this treatment may not be suitable for everyone.

What is emicizumab?

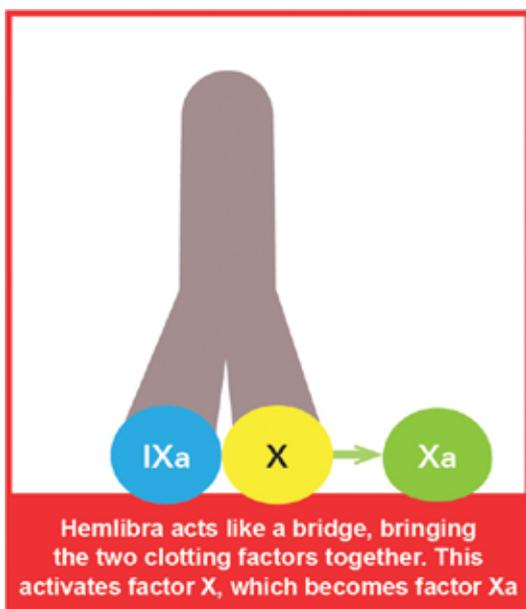
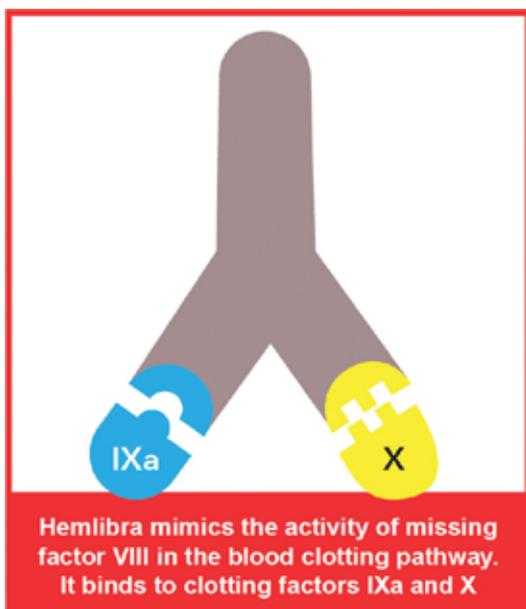
Emicizumab is a treatment called a monoclonal antibody. Monoclonal means single type. So it is a single antibody that has been manufactured in a laboratory in large quantities and designed to do a specific job in the body. It isn't made from blood products so there is no viral infection risk.

Emicizumab copies the way factor VIII works. It brings activated factor IX and factor X together in the blood, which activates factor X. This allows blood clotting to continue and ultimately form a clot and stop bleeding. Emicizumab is present in the blood at stable levels, helping to prevent most bleeds and gives protection against minor injuries.



For everyone affected by a genetic bleeding disorder

To find out more, visit haemophilia.org.uk or contact us on 020 7939 0780 or at info@haemophilia.org.uk



This allows the clotting pathway to continue and the blood to clot

IXa = activated factor nine, X = factor ten, Xa = activated factor ten.

Except in very specific circumstances and under clinical supervision, you cannot use emicizumab with FEIBA.

If your usual bypassing agent to stop bleeding is FEIBA, you will need to switch to rFVIIa (7a).

Use of higher doses of FEIBA with emicizumab can cause potentially life-threatening complications.

If you need high doses of FEIBA, you will have to have these in hospital so you can be monitored.

Because it is a newer treatment, we don't fully understand all the possible risks yet. Emicizumab helps your blood to clot. If the blood clots when it shouldn't, it can cause heart attacks, strokes and thrombosis.

If you are older and have other risk factors for these conditions, or they run in your family, you will need to discuss whether emicizumab is right for you with your haemophilia specialist.

What are the benefits of emicizumab treatment?

You may be able to have fewer injections. Your treatment to prevent bleeds (prophylaxis) will be one subcutaneous injection weekly or fortnightly. You may need additional treatment if you do have a bleed. If you have a chronic inhibitor, emicizumab gives better protection against bleeds than bypassing agents.

Emicizumab can be used to provide protection against bleeds while you are having immune tolerance induction (ITI) treatment for an inhibitor.

For your regular bleed treatment, you may then change back to FVIII or stay on emicizumab. You'll need to discuss with your

Who can have emicizumab?

Emicizumab is a treatment for anyone with:

- severe haemophilia A without inhibitors
- haemophilia A of any severity with inhibitors

haemophilia team which is the better option for you.

Are there any drawbacks to emicizumab treatment?

If your condition is stable on your current treatment and you are happy with your treatment plan and have good vein access there may be less reason to switch, particularly if you don't have many bleeds.

You need specialist blood tests to monitor your factor level in an emergency if you are on emicizumab as normal factor level tests will not show the action of emicizumab. These are not available everywhere, so it may be an issue if you travel a lot.

Emicizumab takes up to 6 months to get out of your system.

How you give emicizumab

Emicizumab is given as a subcutaneous injection. That means an injection you have just under your skin, not into a vein. You give yourself this injection once a week or once a fortnight, depending on your doctor's instructions.

The injection can be given under the skin in different areas of the body, these include upper arm, the tummy, and the thigh.

Injections must be given at least 2.5cm from the site of the previous injection.

What happens when you switch from your current treatment to emicizumab?

When you start on emicizumab, you have one injection of a 'loading dose' each week for 4 weeks. After 4 weeks, your dose of emicizumab will change to a smaller dose if you are having emicizumab weekly. Your dose will stay the same if you're having it fortnightly.

You should have your first loading dose at your haemophilia centre. This is so that the staff can teach you how to give the subcutaneous injections yourself and monitor you for any side effects. Some centres may ask you to attend the centre for all your loading doses.

You must **stop** your prophylaxis with FEIBA or rFVIIa (7a) 24 hours before your first dose of emicizumab. Once you are having emicizumab, unless you have agreed with your specialist, **you must not have any further treatment with FEIBA unless you are under supervision of a doctor.** Using FEIBA alongside emicizumab can potentially cause life threatening complications.

Your centre will give you clear instructions on what to do and if you are unsure always contact them to check before doing anything.

How will I be monitored on emicizumab?

Your centre will aim to keep the number of blood tests you have as low as possible – they'll discuss with you which ones are necessary. You are likely to need blood tests:

- After your loading doses, to monitor levels of emicizumab in your blood
- If you have a bleed and need factor or bypassing agents (FEIBA or rF7a)

You may need additional blood tests if:

- Your bleeding pattern improves but then worsens again – your team will want to check for antibodies against emicizumab (these are rare, but have been reported)
- You have a chronic inhibitor – your team will check your inhibitor levels once or twice a year

What happens if I miss a dose?

If you miss a dose of emicizumab, call your centre. You may be advised to have a dose as soon as you remember, but **do not** have two doses in one day to make up for a missed dose.

What happens if I have a bleed?

Emicizumab prevents bleeds, but it isn't a treatment for bleeding. When you switch to emicizumab, check with your haemophilia team how to manage a bleed or injury – it may be different to what you've done in the past or you may need a lower dose because you are having emicizumab.

In the first few months after starting emicizumab, it is best to contact your treatment centre for advice if you do have a bleed or injury, however minor.

Generally, for minor problems, you can take tranexamic acid and wait to see if that helps. But don't do this without your teams' advice. **You must contact your treatment centre if a single dose of factor or NovoSeven (rFVIIa) is not enough to treat your bleed adequately.**

Apart from your haemophilia medication, treat bleeds as you normally would, with PRICE – protection, rest, elevation and ice. Emicizumab may increase the risk of skin damage when ice is used, so take extra care to make sure that the ice doesn't have direct contact you're your skin and report any skin discoloration or discomfort to your haemophilia centre.

What happens if I need surgery?

If you need surgery, you must ensure your haemophilia centre is involved in the planning of your care. For minor surgery such as simple dental extraction or endoscopy people have been able to have

treatment without additional treatment cover, but this decision must be made by your haemophilia centre.

Will I still have to fill in Haemtrack?

When you start on your emicizumab, it is important that you keep a record of all your treatment and any bleeds on Haemtrack. This helps you and your haemophilia team to monitor how well emicizumab is working.

Are there any common side effects?

The most common side effect with emicizumab is irritation where you give the injection. This happens in around 1 in 5 people. You may have redness, soreness or itching or the area may feel hard. To avoid injection reactions, don't give two injections running in the same place.

Other common side effects are headache and aching joints.

Tell your treatment centre if you have unusual headaches, joint pain or other new symptoms.

Are there any serious side effects?

Two potentially serious side effects have been reported in people having emicizumab. These are blood clots (thrombosis) and an unusual clotting condition called TMA or thrombotic microangiopathy.

Blood clots (thrombosis) can cause problems including heart attacks or strokes. They could happen to anyone taking emicizumab but are probably more likely in older people who have other risk factors for thrombosis. Contact your doctor immediately if you have any of these symptoms:

- chest pain or tightness; shortness of breath or coughing blood
- confusion

- eye pain or swelling or difficulty seeing
- fast heart rate
- feel faint
- headache
- numbness in your face
- pain or redness in your arms or legs
- weakness in any part of your body

TMA means there are small blood clots and damage in the tiniest blood vessels in the body. These can harm your kidneys, brain and other organs.

This has only happened in patients with inhibitors on emicizumab who had FEIBA at higher doses to treat bleeding. You must not give yourself any FEIBA without your doctor's agreement.

If you can't have rFVIIa (7a), your doctor will advise you on what to do if you do have a bleed.

Contact your doctor immediately if you have any of these symptoms:

- Weakness
- Swelling of arms and legs
- Confusion
- Yellowing of skin and eyes
- Stomach, abdomen or back pain
- Feeling sick or vomiting
- Decreased urination

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