Extended half-life (EHL) factor VIII

- A new form of factor VIII is becoming available to treat severe haemophilia A
- Extended half-life factor can stay in the bloodstream for longer, so you can have injections to prevent bleeds (prophylaxis) less often
- Bleeds can generally be controlled with a single dose
- There is no infection risk as the products are entirely lab-made
- There appear to be few side effects
- The products seem no more likely to cause inhibitors than regular factor VIII
- They may not be suitable for everyone – you need to discuss your own personal bleeding and treatment history with your doctor and have blood tests before and after a trial dose

Who can have EHL factor VIII?

These drugs have been developed for anyone being treated for severe haemophilia A. The aim is to keep your lowest ('trough') factor VIII levels above 1%. In practice, you would have to discuss your medical and bleed history with your own specialist in order to decide whether EHL factor is a good option for you.

How often you have treatment depends on blood test results, your history of bleeds and on how active you normally are. In trials, patients have been able to reduce their prophylaxis injections to around once or twice weekly. Those who play sports at weekends may be able to adjust their dose timing to best cover those activities. Those who are least active may be able to have treatment only once a week.

‘On the EHL trial I’ve had factor injections every five days instead of every other day. It doesn’t sound much, but it’s really freed me up. I don’t have to plan as much and I don’t have to worry about finding somewhere private to give an injection.’ Ed, haemophilia A patient

Children under 12 are likely to need injections more often as factor VIII generally doesn’t last as long in their bloodstream. They may be able to reduce the dose and/or frequency of injections as they get older.

Research is still looking into the best way to use these products in very young children. It’s not yet clear whether doctors should start previously untreated patients on EHL factor VIII or regular factor VIII.

What happens when you switch from regular to EHL factor VIII

Once you’ve agreed with your specialist to try EHL factor VIII the first step is to have a test dose. Your specialist will need to check your factor VIII levels before the test dose and then 15 minutes, 6 hours, 24 hours, two days and three days afterwards.

This fact sheet is about a new form of factor VIII that can stay in the bloodstream for longer. It is called extended half-life factor, or EHL. If this treatment is suitable for you, you may be able to have injections to prevent bleeds (prophylaxis) less often and are likely to need fewer injections to control any bleeds you do have.

There are several types of EHL factor VIII in development but currently only one licensed, called Elocta.

What is EHL factor VIII?

EHL stands for ‘extended half-life’ although it is occasionally called ‘enhanced half-life’. It means that the factor can stay in the blood for longer. For many patients, this means being able to have factor VIII injections less often.

These drugs are entirely made in the laboratory (they are ‘recombinant’). They don’t come from human blood plasma so there’s no infection risk.

The first available form of EHL factor VIII is called Elocta. It is a ‘recombinant factor VIII Fc fusion protein’. The factor is fused to a section of protein (called Fc) taken from a human antibody (immunoglobulin). This fusion protein is made inside lab-grown human cells.

The antibody protein in the drug enables it to circulate in the blood for longer. In theory, using human cells to manufacture the drug means that it is less likely to cause inhibitors to form, but these drugs are too new for us to know that for certain. The new drugs haven’t yet been tested in patients who already have inhibitors.

In Elocta trials, some patients had weekly prophylaxis injections but most had individually worked out treatment plans, with injections every three to five days. By the end of the trial, 1 in 3 adults on an individually worked out treatment plan were able to have injections every five days. Nearly half of adults and children had no bleeds during the study.
You then have another appointment with your specialist to agree your dose, how often you have injections, what you should do if you have a bleed and when you need to contact your haemophilia centre.

After you’ve started EHL factor you need to have follow-up appointments every four weeks for three months (some of these may be over the phone). After 10 treatment days you will have blood tests for inhibitors. (For example, if you have injections every four days this will be after 40 days). After three months you will have a final appointment to review how you are getting on, have peak and trough factor VIII levels measured and more tests for inhibitors.

Your dose and how often you have the injections may change, depending on your blood test results and whether you have any spontaneous bleeds. If you’re having bleeds related to sports or other activity, talk to your doctor about adjusting the timing of your treatment so your factor level is highest when you’re most active. If you haven’t had any bleeds you may be able to have injections less often.

In trials, nearly 8 out of 10 patients increased or maintained their activity levels while on the new treatment without having any more bleeds.

**What you have to do**

When you start on your new factor VIII it’s important that you keep a record of when you have your injections and of any bleeds. To be prescribed EHL factor you have to agree to do this using Haemtrack on line, on paper or using the Haemtrack app. You will also need to have regular blood tests to check your factor trough levels and test for inhibitors.

*The key things this new treatment has given me are confidence and reassurance. I feel I can be more active, stay out for longer and go on holiday with less worry.*

Steve, haemophilia A patient

**What happens if you have a bleed**

In trials, a single dose of EHL factor VIII has generally been enough to control bleeds. Until you have experience managing your new treatment your haemophilia centre will ask you to let them know if you have a bleed and contact them again if the bleed isn’t controlled within 24 to 48 hours. If you consistently need more than two factor doses to control a bleed your doctor will re-check your peak and trough factor levels.

From trials, doctors have found that breakthrough bleeds are most likely to be prevented by increasing how often you have the injections but keeping to the same overall total dose.

**Side effects**

It’s too soon to be definite, but so far the EHL factor VIII seems no more likely to cause inhibitors to develop than regular factor VIII. No patients on the trials have developed inhibitors.

As with many drugs, there is a long list of possible side effects. Many of these are symptoms that everyone is likely to have at some time, such as cold and flu symptoms, and are unlikely to have been caused by the drug. Apart from these, the commonest side effects in the adult trial of Elocta (in patients on individual treatment plans) were:

- joint pain in 9 out of every 100 (9%)
- headache in 4 out of every 100 (4%)
- raised temperature in 3 out of every 100 (3%).

In the children’s trial, the only side effects thought to be caused by Elocta were muscle pain (in 4%) and red rash (in less than 1%).

It’s possible to have an allergic reaction to any factor VIII product. This could cause itchy rash, tightness in the chest, wheezing, a fall in blood pressure and collapse. No reactions like this have been seen so far in trials for Elocta.

**About The Haemophilia Society**

We are the only UK wide charity for everyone affected by a bleeding disorder; a community of individuals and families, healthcare professionals and supporters.

More than 25,000 men, women and children in the UK have a diagnosed bleeding disorder, and the number rises every year. Membership of The Haemophilia Society is free and open to all.

To find out more, or to become a member for free, visit our website at haemophilia.org.uk or call us on 020 7939 0780.

The Haemophilia Society makes every effort to make sure that its services provide up-to-date, unbiased and accurate information about bleeding disorders. We hope that this information will add to the medical advice you have received and help you to take part in decisions related to your treatment and care. Please do continue to talk to your doctor or specialist nurse if you are worried about any medical issues.

**Give us your feedback**

We hope you have found this information helpful. If you have any comments or suggestions about this factsheet or any of our other information, please write to the Head of Membership and Planning at the address below.

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