**Extended half-life (EHL) factor IX**

- A new form of factor IX is becoming available to treat severe haemophilia B
- 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 IX

This fact sheet is about a new form of factor IX 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 a few different types of EHL factor IX that are made in different ways. Currently there are two that are licensed, called Alprolix and Idelvion.

**What is EHL factor IX?**

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 IX injections less often.

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

Alprolix is a recombinant factor IX 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.

Idelvion is made in a similar way, but using a technique called albumin fusion. The factor IX is fused to lab-made human blood protein (albumin). The fusion protein is also made inside lab-grown cells, in this case called ‘Chinese hamster ovary’ cells or CHO.

It is the way these drugs are made that enables them to circulate in the blood for longer. In theory, using human proteins to manufacture the drugs means that they are 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 clinical trials for Alprolix and Idelvion, some patients had prophylaxis injections once a week and some had individually worked out treatment plans, with injections every 10 to 14 days. Around 1 in 4 of those having weekly injections had no bleeds during the study. Of those having individually worked out treatment plans, around 1 in 2 had no bleeds.

**Who can have EHL factor IX?**

These drugs have been developed for anyone being treated for severe haemophilia B. The aim is to keep your lowest (‘trough’) factor IX levels at between 1% and 3%. 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 how active you normally are. In trials, patients have been able to reduce their prophylaxis injections to between once a week and once a fortnight.

Those who play sports at weekends may be able to adjust their dose timing to best cover those activities. You would need to discuss this with your specialist first.

Children under 12 are likely to need injections more often as factor IX 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.

‘If my son hadn’t been on the trial he’d be having prophylaxis every three days. Now he only has one injection a week, so the new drug has made a huge difference, more than halving the injections he needs.’ Sara-Jane, mum of Henry, a haemophilia B patient

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 IX or regular factor IX.
What happens when you switch from regular to EHL factor IX

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

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 seven days, this will be after 70 days). After three months, you will have a final appointment to review how you are getting on, have peak and trough factor IX 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 haven’t had any bleeds, you may be able to have injections less often. If you are 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 are most active.

What you have to do

When you start on your new factor IX, it is 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.

‘Because I already had joint damage, the EHL hasn’t enabled me to do specific things that I couldn’t do before. But I can stay out for longer and do more with my family. I’m more confident about going out and so is my wife.’ Dave, a haemophilia B patient

What happens if you have a bleed

In trials, one or two doses of EHL factor IX have 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.

Side effects

It’s too soon to be definite, but so far the EHL factor IX seems no more likely to cause inhibitors to develop than regular factor IX. 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 trials for both drugs, (for patients on individual treatment plans) were:

- headache
- injection site reaction
- joint pain.

It’s possible to have a serious allergic reaction to any factor IX 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 these new extended half-life factor IX drugs.

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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