THE HAEMOPHILIA SOCIETY

FACTSHEET

Extended half-life (EHL) factor VIII

This fact sheet is about extended half-life treatment which is now available for all people with haemophilia A in the UK. These are factor VIII products that can stay in the bloodstream for longer. If this treatment is suitable for you, you may be able to have injections to prevent bleeds (prophylaxis) less often, or may stay on the same dose/ schedule and have higher trough levels. You are likely to need fewer injections to control any bleeds you do have.

Previously many people living with haemophilia A had restricted access to EHL however now there are several types of EHL factor VIII in development.

There are three currently licensed in the UK

Elocta

- Elocta is manufactured and marketed by Sobi
- It is used to control and prevent bleeding in people with haemophilia A (congenital factor VIII deficiency) in all age groups.
- Elocta is a 'recombinant factor VIII Fc fusion protein'. The factor is fused to a section of proten (called Fc) taken from a human antibody (immunoglobulin). This fusion protein is made inside lab-grown human cells.
- Elocta can also be used on demand, meaning it is administered to help stop a bleed as it is occurring.

Esperoct

- Esperoct® is manufactured and marketed by Novo Nordisk
- It is used to control and prevent bleeding in people with haemophilia A (congenital factor VIII deficiency) in people of 12 years and above.
- Esperoct® can also be used on demand, meaning it is administered to help stop a bleed as it is occurring.
- Esperoct® acts as a replacement for your missing or deficient clotting factor VIII. It is made up of a recombinant factor VIII molecule and an attached PEG (polyethylene glycol) molecule.

Adynovi

- Adynovi is manufactured and marketed by Takeda
- It is used to control and prevent bleeding in people with haemophilia A (congenital factor VIII deficiency) in people of 12 years and above.

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

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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 people, this means being able to have factor VIII injections less often or being able to have higher trough levels (trough is the level of factor in the blood just before the next infusion).

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

It is the way these drugs are made that enables them to circulate in the blood for longer. The factor VIII molecule is attached to another molecule (Fc or PEG) which makes it more stable in the blood.

These drugs are too new for us to know how they affect the risk of developing inhibitors the studies in people who have had lots of factor VIII before show no increased inhibitor rate; studies in children with low/no exposure to factor VIII before starting EHL are on-going.

Who can have EHL factor VIII?

These drugs have been developed for anyone being treated for 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 haemophilia team 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.

Children under 12 are likely to need injections more often, or higher doses than adults 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.

What happens when you switch from regular to EHL factor VIII?

Once you've agreed with your haemophilia team to try EHL factor VIII the first step is to have a test dose, your factor VIII levels will be taken before the test dose and then 15 minutes, 6 hours, 24 hours, two days and three days afterwards.

You then have another appointment 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 10 treatment days you might have a blood test to check for an inhibitor (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 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 previous 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 online, 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.

What happens if you have a bleed

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.

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

Treatment of bleeding

The dose of EHL is calculated depending on your body weight and the factor VIII levels to be achieved.

Side effects

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.

It's possible to have an allergic reaction to any factor VIII product. Signs of an allergic reaction include: wheezing, chest tightness; Redness and/or swelling of the lips, tongue, face or hands; rash, hives, weals or itching; pale and cold skin, fast heartbeat, or dizziness (low blood pressure; headache, nausea or vomiting; difficulty in swallowing or breathing

If you think that you have experienced any side effect, you should contact your haemophilia centre immediately.