Introduction

This leaflet aims to provide you with general information about children with Severe Haemophilia A and an Inhibitor. If you are ever worried about your child please contact your IBD Team or take them to the Accident and Emergency Department.

What are inhibitors?

How are they managed in severe haemophilia A

Inhibitors are antibodies. Antibodies are one of the ways in which the body tries to protect itself from harm. They are made in response to foreign proteins (antigens). The antibody locks onto the antigen to neutralise its effect. This neutralising effect is helpful when controlling such things as infection, but less good when the body is in need of an injected protein such as Factor 8.

Approximately 20% of children with haemophilia A develop inhibitors to Factor 8, usually between the eighth and fourteenth treatment.

How are inhibitors detected?

At each clinic appointment we routinely take blood to check for the presence of inhibitors. The signs and symptoms of inhibitors may:

- only be noticeable on blood tests
- be undetectable on blood tests, but bleeds need more or higher doses of Factor 8 than expected to stop the bleeding.
- be obvious on blood tests as well as there being increased bruising and bleeding into muscles and joints that do not respond to factor 8 replacement. These are known as symptomatic Inhibitors

Inhibitors can make managing haemophilia difficult so we aim to put a new treatment plan in place quickly. We want to reduce painful bleeding episodes and minimise the risk of joint and muscle damage, helping to get life with haemophilia back on track.

Managing symptomatic inhibitors

If we find your son has an inhibitor we will let you know. We will repeat the blood tests and ask you some questions about any change in symptoms, such as increased bruising or spontaneous bleeds. Once we have confirmed that your son has an inhibitor we will talk to you about current treatment.
First line management of inhibitors has recently changed. In the past we "swamped" the inhibitor with high doses of Factor 8 (immune modulation), but recent treatment developments have provided us with an effective alternative option in the form of a manufactured therapeutic antibody, Hemlibra (Emicizumab).

Emicizumab does not get rid of inhibitors but bypasses them, reducing the amount of damage they cause.

Emicizumab is given under the skin (subcutaneously) once a week. It reduces the risk of spontaneous joint and muscle bleeds, but does not prevent all bleeding episodes.

It should not be used if your son has an injury or you think he might have a bleed. These episodes will need to be managed with tranexamic acid or an injection of Factor 7a. We will make sure that you have a supply of both medicines to keep at home.

**Giving Emicizumab**

We will take time to help you learn how to give treatment. We do most of the training in the Haematology Treatment Room (HTR) but we know that giving treatment at home is different to the hospital environment, so we will also come to your home to help you to start giving treatment there. We will also help you to find the best place to store treatment and equipment.

We prefer there to be more than one person who is able to give your child treatment. The second person does not have to be a family member, but it does need to be someone you can trust.

**Self-treatment**

We like to teach children to treat themselves as early as possible, and definitely before they start high school. Children learn as they watch you at home, and us in the HTR. We do 1:1 training and may also give group training, which is a good place for the children to share their tips.

**If your son has injured himself or you think he has a bleed:**

*Use usual first aid first! - RICE - Do not give Emicizumab.*

**Rest** - Rest the affected joint or muscle. If you son has an elbow or shoulder bleed he should rest his arm and not use it to lift or carry things. If it is a joint or muscle bleed in his leg he should not walk but rest as much as possible.

**Ice** - Apply ice, wrapped in a towel, or a cold pack to the affected joint, either by using an ice pack, crushed ice in plastic bag or frozen vegetables. Apply for 10-15 minutes. Ice helps to reduce pain and muscle spasms as well as reducing swelling and redness. Ice alternating with heat can also help to reduce pain, particularly when the bleed is into a muscle.

**Compression** - Applying pressure to the injured area will help slow blood flow; the pressure can also provide comfort. A Tubigrip bandage is a good example of compression and can be used over the knee, ankle, elbow or wrist. Compression is often too painful in the early stages of a bleed.

**Elevation** - Raise the injured arm or leg above the heart although this may seem difficult you can prop an arm and leg up with cushions or pillows. This helps to reduce the blood flow to the area and lower blood congestion to slow the bleeding.

If the symptoms of a possible bleed continue please contact the IBD team for advice. Where they are unavailable please go straight to the accident and emergency department taking one of your two doses of emergency Factor 7a with you.
Links
The Haemophilia Society website: https://haemophilia.org.uk/
Alder Hey Children’s NHS Foundation Trust: https://alderhey.nhs.uk/

Contact Information Haematology Nurses 8.30am-5.00pm 0151 252 5070 with answer machine. Queries will be reviewed by the end of the next working day.

Nicki Mackett: 07584 234 526 nicki.mackett@nhs.net
Catherine Benfield: 07876 132 163 catherine.benfield@nhs.net

This leaflet only gives general information. You must always discuss the individual treatment of your child with the appropriate member of staff. Do not rely on this leaflet alone for information about your child’s treatment.

This information can be made available in other languages and formats if requested.

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