Acquired Haemophilia A

This leaflet offers more information about acquired haemophilia A. If you have any further questions or concerns, please speak to the staff member in charge of your care.

What is acquired haemophilia A and why have I got it?

Acquired haemophilia A is a bleeding disorder which develops suddenly unlike congenital haemophilia with which people are born. In acquired haemophilia A your immune system has started to neutralise the factor VIII (eight) produced by your body. We call this an inhibitor. Factor VIII is one of the clotting factors that our body needs to help stop bleeding when it occurs. People with acquired haemophilia A will bleed and bruise more than the average person, often to a dangerous extent. This is a serious condition that requires urgent hospital admission for treatment and management. It is likely you will need to stay in hospital for four to six weeks.
The incidence of acquired haemophilia A is approximately 1.5 in 1 million.

For about 50% of people with acquired haemophilia, no underlying cause will ever be found. Autoimmune disorders, cancer, pregnancy and drug reactions have been associated with acquired haemophilia. As part of your treatment you may have tests, including scans, to rule out these conditions as a cause.

What are the signs and symptoms?

- Bruising is the most common symptom in acquired haemophilia and you may have developed massive bruises without any recollection of injuring yourself.
- Blood in your stool or in your urine.
- Prolonged clotting screen.

It is important to remember that acquired haemophilia is very rare and doctors who are inexperienced with this condition may not diagnose it immediately. Approximately 35% of patients with acquired haemophilia go undiagnosed for more than seven days.

Do I need any tests to confirm the diagnosis?

The main tests we will do to diagnose and monitor your condition are blood tests checking your factor VIII and Bethesda levels. Normal factor VIII levels are 50-150i.u./dL, and it is not uncommon for patients with acquired haemophilia to have a factor VIII of less than 10i.u./dL or even an undetectable factor VIII.

The Bethesda test measures the level of inhibitor working against the factor VIII in your body. It will take a number of weeks for this level to reduce with treatment.

Another test we run regularly is your full blood count (FBC). We will be paying particular attention to your haemoglobin (Hb). Haemoglobin is a protein in your blood that carries oxygen around your body. If that number drops suddenly it may be a sign you are bleeding.

What treatments are available?

The goal of treatment in acquired haemophilia is to control bleeding and eradicate the inhibitor. If you are bleeding we may commence you on a bypassing agent. Bypassing agents are products we give you through your vein and they contain clotting factor(s), other than factor VIII, in high amounts. This works by supercharging your body's clotting system so it can work without factor VIII. You will be given further information on these products if they are offered to you. **It is important to tell your doctor or nurse if you decline blood products on religious grounds.**

If you need to start on a bypassing agent we may also wish to insert a Peripherally Inserted Central Catheter (PICC). This is to reduce how often we need to insert needles into your arm and reduce your risk of bleeding and bruising as a result of this. If we offer you a PICC line we will give you additional patient information on this.

To eradicate the inhibitor we need to suppress your immune system. We do this by giving you high dose steroids. We may also give you medication called rituximab or cyclophosphamide. We have separate patient information leaflets on these drugs. Side effects of high dose steroids include increased susceptibility to infection, increased appetite, weight gain, fluid retention, irritability, mood swings, confusion, delirium, difficulty sleeping and elevated blood sugars.

What happens if I do not get treatment?

If you decide to decline the advised treatment you may suffer from a life- or a limb-threatening bleed.

Is there anything I can do to help myself?

- Suppressing your immune system also makes you more prone to infection and this may be life-threatening. To help prevent this we give you a variety of antimicrobial drugs. It is important that you take these tablets as prescribed and do not stop treatment until you are told. Contact the haemophilia office if you need your prescription refilled.

- Contact the hospital immediately if you have a temperature above 37.5 degrees Celsius or if you feel generally unwell. Avoid people who are unwell and follow NHS guidelines on avoiding food poisoning. Please also

inform us if you are exposed to anyone with chicken pox or shingles as we may need to give you extra treatment to protect you.

- When you are discharged from hospital you will still need to attend the hospital regularly for blood tests. You may need to attend twice weekly initially and weekly thereafter for a number of months. It is important to attend these appointments so we can monitor your condition and wean you off steroids safely. Stopping high-dose steroids without an appropriate weaning regimen can be dangerous, so please follow our guidance closely.

- There is nothing you have done to cause the acquired haemophilia and there are no special foods or supplements you can take, other than those we have prescribed, that will effectively treat this condition. Please discuss all new over-the-counter medication or herbal supplements with the haemophilia team while you are being actively treated. Please discuss any proposed vaccinations or invasive treatments with the haemophilia team.

Useful sources of information

https://rarediseases.org/rare-diseases/acquired-hemophilia/

https://www.nhs.uk/live-well/eat-well/10-ways-to-prevent-food-poisoning/

This leaflet includes information obtained from the European Acquired Haemophilia Registry (EACH2) and Hays, C.R.M. (2013) 'Acquired Disorders of Coagulation: The Immune Coagulopathies' in Marder, V.J. et al. (ed.) *Haemostasis and Thrombosis Basic Principles and Clinical Practice*, Philadelphia: LWW pp723-730.

Contact us

If you have any questions or concerns about acquired haemophilia, please contact the haemophilia office on 020 8725 0763 (Monday to Friday, 8am to 5pm). Out of hours, please contact the haematology registrar via the hospital switchboard.

If your query is in regards to your appointment for your blood test please call the Haematology Day Unit on 020 8725 1680.

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.stgeorges.nhs.uk

Additional services

Patient Advice and Liaison Service (PALS)

PALS can offer you on-the-spot advice and information when you have comments or concerns about our services or the care you have received. You can visit the PALS office between 9.30am and 4.30pm, Monday to Friday in the main corridor between Grosvenor and Lanesborough wings (near the lift foyer).
Tel: 020 8725 2453 **Email:** pals@stgeorges.nhs.uk

NHS Choices

NHS Choices provides online information and guidance on all aspects of health and healthcare, to help you make decisions about your health.
Web: www.nhs.uk

NHS 111

You can call 111 when you need medical help fast but it's not a 999 emergency. NHS 111 is available 24 hours a day, 365 days a year. Calls are free from landlines and mobile phones.
Tel: 111

AccessAble

You can download accessibility guides for all of our services by searching 'St George's Hospital' on the AccessAble website (www.accessable.co.uk). The guides are designed to ensure everyone – including those with accessibility needs – can access our hospital and community sites with confidence.

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