

Automated red cell exchange for sickle cell disease

Information for children, young people and their parents



This information sheet/leaflet explains a procedure used to treat sickle cell disease called automated red cell exchange. It includes the risks, the benefits and the alternatives. If you have any questions or concerns, please do not hesitate to speak to any member of the team caring for you.

Confirming your identity

Before you have a treatment or procedure, our staff will ask you your **name** and **date of birth** and check your **ID band**. If you don't have an ID band we will also ask you to confirm your address.

If we don't ask these questions, then please ask us to check.

Ensuring your safety is our primary concern.

What is automated red cell exchange?

It is a type of blood transfusion where your red cells are removed with the help of a cell separating device called an apheresis machine and then replaced with non-sickle red cells from a donor. The apheresis machine enables us to closely balance the amount of blood and fluids entering and leaving your body and ensure your blood pressure and other vital signs are stable.

To have this procedure you need to have two good veins, usually in your arms, where we can put in two cannulas (thin plastic tubes) or one cannula and one needle: one to remove sickled blood and one to transfuse the donor red blood cells to you.

This means it can sometimes be difficult to give you this treatment as a child because you are more likely to have small/difficult veins and will need to discuss other options available for us to be able to carry out the automated exchange.

Why do I need automated red cell exchange?

In an emergency – you may be given this treatment to help improve symptoms such as:

- a severe sickling episode affecting your lungs (acute chest syndrome)
- an acute stroke affecting your brain.

As planned treatment – you may be given this treatment:

- as part of a long-term programme to prevent stroke or other sickle complications
- if you have a number of painful sickling episodes affecting your lungs (acute chest syndrome) and hydroxycarbamide (hydroxyurea) has not helped.
- if you have frequent and disruptive episodes of acute pain and hydroxycarbamide (hydroxyurea) has not worked/helped these
- to prepare you for an operation.



What are the risks?

Minor reactions

Hives, rash or a mild fever. You can treat these by taking an antihistamine and paracetamol.

Iron overload

This is common if you have regular top-up blood transfusions. Excess iron can be removed with medication (daily tablets or injections). You are less likely to have iron overload if you have automated red cell exchange.

Antibodies

We match your blood very closely with the donated blood. But you can still develop antibodies against donor blood, which means we have to do further matching to prevent you having a delayed transfusion reaction. Developing antibodies can mean it takes much longer to find suitable blood for exchange. If you develop them, we will send you an alert card that says 'I need special blood'.

You must keep this card on you at all times in case you need a transfusion and show it to any and all health professionals who treat you.

Delayed transfusion reaction

You can have a delayed transfusion reaction where your body abnormally breaks down the blood that has been transfused. These red cell antibodies could be newly formed or too weak to pick up in laboratory tests. This may cause:

- severe general pain
- blood in your urine or you urine may be red or cola coloured
- fatigue
- shortness of breath
- fever
- pain in your loin or back.



If you have any of these symptoms, please go straight to your local Emergency Department (ED/A&E) and tell the medical staff that you have recently had a transfusion.

Low blood calcium

You can sometimes have low blood calcium during the procedure. This makes you feel light headed, dizzy or faint, or you may have a tingling feeling in your lips and fingers. If this happens we will give you some calcium tablets/intravenous calcium during the procedure or extra fluid to reduce the symptoms.

Consent

We must by law obtain your written consent to any operation and some other procedures beforehand. Staff will explain the risks, benefits and alternatives before they ask you to sign a consent form. If you are unsure about any aspect of the treatment proposed, please do not hesitate to ask to speak with a senior member of the staff again. We will ask for your consent for the automated red cell exchange every year.

Low platelet count

Your platelet count will drop straight after the procedure. This will not cause any problems and your platelet count will go back to normal within 24 hours.

What are the benefits?

- It is quicker, safer and a lot more efficient than a manual exchange transfusion in rapidly reducing the number of sickle blood cells in your body and reducing your iron burden.
- It can be used to rapidly cut the number of sickle cells in your blood when you are severely unwell with, for example, acute chest syndrome or stroke, to quickly reduce the damage caused by sickling.



- It can help to reduce iron build-up in your body. Some patients on automated red cell exchange programmes do not need to take extra medicines to remove iron from their bodies
- It can be quicker than a simple top-up transfusion if you are on a long-term transfusion programme.

Are there any alternatives?

Alternative treatment would include a manual exchange, or a simple top up transfusion. These would need to be discussed further with your doctor as the treatment option would vary depending on the reason for needing it.

Do I need to prepare?

We will send you an appointment for a blood test, which you will have one or two days before the procedure. This enables us to cross-match donor units ready for the transfusion.

Please make sure you drink plenty of fluid before you have the transfusion to make it easier for us to find a vein.

What happens before the procedure?

When you come to the ward, we will take your height and weight and measure your blood pressure, heart rate, oxygen saturation and temperature.

What happens during the procedure?

The apheresis nurse who is doing your red cell exchange will ask you to sit upright in a chair or bed. They will put two cannulas (thin plastic tubes) into veins in your arms: one to remove your blood and one to transfuse the donor blood into you. These will be connected to the apheresis machine.



You will be able to read, watch a DVD, eat and drink while having the transfusion.

How long does it take?

It depends on your height, weight and how much blood you need. The procedure itself takes 2-3 hours, but the appointment is a full day appointment.

What happens after the procedure?

The nurse will monitor you closely on the ward. They will do a blood test to check your full blood count and calcium levels as well as how many sickle red cells you have left in your blood.

You should also be given your next appointment before you leave the ward.

How to I cancel or change my appointment?

Please contact the clinical nurse specialists:

Clinical nurse specialist

Tel: **020 3299 4752** or **020 3299 1916**

Who can I contact with queries or concerns?

If you have any questions, please do not hesitate to contact the paediatric sickle cell team.

Phillip Isaacs Ward (day care)

Tel: **020 3299 4200**

Clinical nurse specialist

Tel: **020 3299 4752** or **020 3299 1916**

Haematology secretary

020 3299 3773

You can call these numbers 9am – 5pm, Monday to Friday



Sharing your information

We have teamed up with Guy's and St Thomas' Hospitals in a partnership known as King's Health Partners Academic Health Sciences Centre. We are working together to give our patients the best possible care, so you might find we invite you for appointments at Guy's or St Thomas'. To make sure everyone you meet always has the most up-to-date information about your health, we may share information about you between the hospitals.

Care provided by students

We provide clinical training where our students get practical experience by treating patients. Please tell your doctor or nurse if you do not want students to be involved in your care. Your treatment will not be affected by your decision.

PALS

The Patient Advice and Liaison Service (PALS) is a service that offers support, information and assistance to patients, relatives and visitors. They can also provide help and advice if you have a concern or complaint that staff have not been able to resolve for you. They can also pass on praise or thanks to our teams. The PALS office is located on the ground floor of the Hambleton Wing, near the main entrance on Bessemer Road - staff will be happy to direct you.

PALS at King's College Hospital, Denmark Hill, London SE5 9RS

Tel: **020 3299 3601**

Email: **kch-tr.palsdh@nhs.net**

If you would like the information in this leaflet in a different language or format, please contact our Communications and Interpreting telephone line on 020 3299 4826 or email kch-tr.accessibility@nhs.net



