UNDERSTANDING SICKLE CELL

What is sickle cell disease?

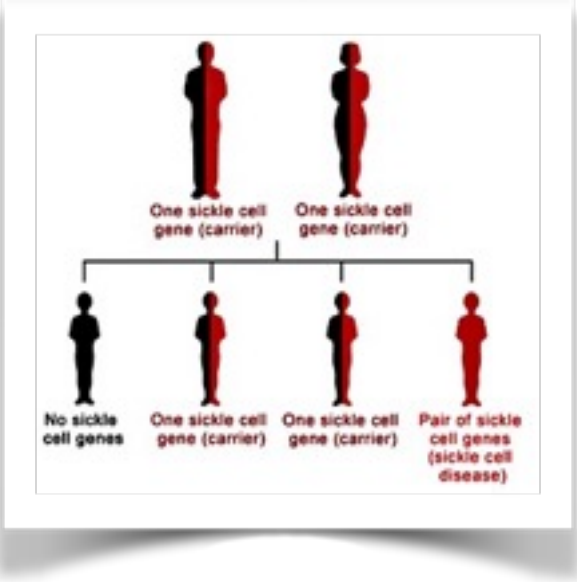
Sickle cell disease (SCD) is by far the most common inherited disease in the world. SCD is a disorder of the blood caused by an inherited abnormal haemoglobin (the oxygen-carrying protein within the red blood cells). The abnormal haemoglobin causes the red blood cells to be distorted. Instead of being shaped like an oval, they are sort of 'S' shaped, a bit like a sickle. These unusually shaped cells can cause problems because they don't live as long as healthy blood cells and they can become stuck in blood vessels.

SCD is a serious and lifelong condition, with sufferers usually experiencing problems from early childhood. They are likely to have anaemia, which is where the red blood cells can't carry enough oxygen around the body, causing the person to feel tired and have shortness of breath. Sufferers also have an increased risk of serious infections and debilitating stroke. Many experience episodes of excruciating pain in various parts of their body. These "sickle cell crises" can be very severe and can last up to a week.

The disease is usually found in people of African heritage. In fact, three out of four cases globally, occur in Africa. Nigeria has the largest burden of SCD in the whole world, with 1 in 50 newborn babies having the disease. It is only due to recent improvements in medical care that affected children have been able to survive beyond childhood. In the UK, 90% of patients with sickle cell disease are of African heritage, although people from the Caribbean, Middle East and Mediterranean regions can have it too.

How is it inherited?

SCD is genetic - it's passed down from parents to children. Children inherit genes from both parents.



- If one parent has sickle cell trait and the other does not, a child could inherit the gene from the parent who has it and pass it on to their own kids.
- If a child inherits only one sickle gene, he is said to be a carrier for the condition, or to have sickle cell trait. A carrier does not develop the disease.
- When both parents have sickle cell trait, their child might be born with sickle cell disease.
- If one parent has the trait and the other parent has the disease, it's quite likely that their child will have sickle cell disease. In this situation, about half of all babies are born with the disease.
- When both parents have sickle cell disease, it's very likely their child will too.

Why do people with SCD need blood transfusions?

A blood transfusion can form part of effective treatment and provide pain-relief for sufferers of SCD. In fact, people with the disease are one of the largest patient groups requiring blood transfusions.

There are two different kinds of blood transfusion treatment used for SCD patients: simple transfusions and exchange transfusions.

- Simple transfusions are used to prevent stroke and brain damage. Typically given in intervals, possibly once or twice a month, they deliver additional healthy red blood cells to the patient's body. A few units of blood are given through a small tube (drip), usually placed in a vein through the patient's arm.
- Exchange transfusions are occasionally needed in the management of a sickle cell crisis. The procedure involves slowly removing the patient's blood and replacing it with blood from a healthy donor. This allows the blood cells to flow more freely, easing disease symptoms and severe pain.

Why are black blood donors so important?

Most patients that need a blood transfusion usually only require a single blood transfusion in their lifetime. They can be given blood that is matched to their ABO and Rh blood groups: A-, B-, AB-, O-, A+, B+, AB+, or O+.

But when a patient has a medical condition like SCD that usually requires multiple blood transfusions, sometimes for the rest of their lives, they have to be given blood that is more extensively matched.

As blood group is linked to a person's genes, SCD patients are more likely to find closer blood matches with people with the same genetic heritage as them. For instance, blood subtype 'Ro' is rare among white European people - only 2% of regular blood donors in England have the Ro subtype. But is found in more than 10 times as many black African and Afro-Caribbean people.

As it's mainly black people with Ro blood and mainly black people with SCD, to treat SCD, we need black people to donate blood. As people with SCD often require blood transfusions throughout their lives, we need black people to donate often and donate for the long term.

Action On Blood is a membership organisation of voluntary blood and organ donors. We believe that no country, community or ethnic group should be left behind in having access to sufficient and safe blood and organ supplies for their health needs. We also believe that true social transformation can only be achieved through the active involvement and support of the communities affected.

Our mission is to build cultures of voluntary donation in communities around the world and support local transfusion and transplant services to deliver universal access to lifesaving blood and organs. We achieve this by empowering individuals to transform their communities from within. Embedded in their respective communities, they have the power to navigate social nuances, challenge negative behaviours and break down attitudinal barriers.

If you would like to transform attitudes to blood and organ donation in your community, contact Action On Blood on actiononblood@gmail.com.