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For more information

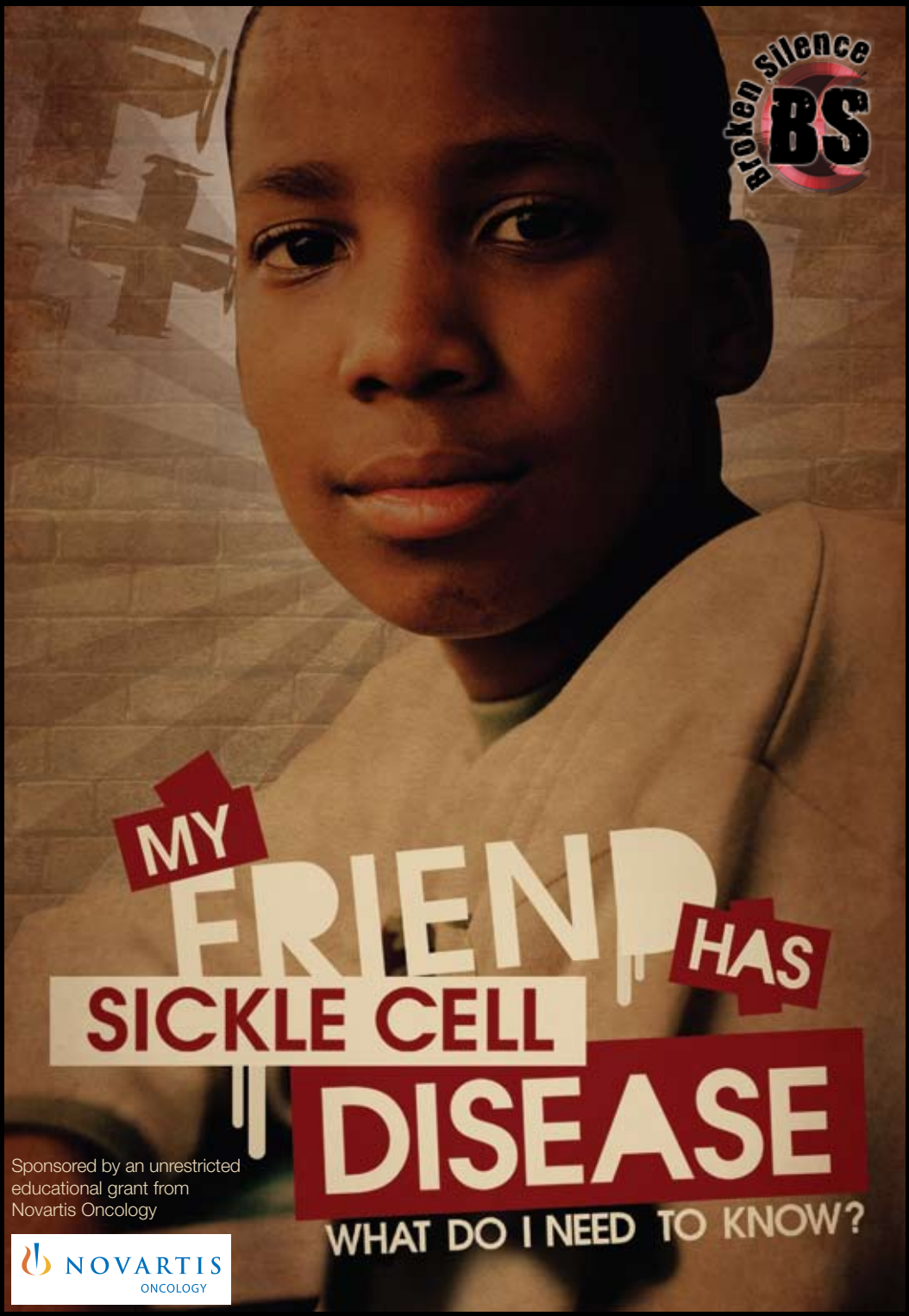
For more information on sickle cell disease or Broken Silence visit www.brokensilence.org or contact Broken Silence via e-mail – brokensilence@brokensilence.org

Useful websites

1. http://www.brokensilence.org/
2. http://www.nhs.uk/Conditions/Sickle-cell-anaemia/Pages
3. www.sickle-thal.nwlh.nhs.uk
4. www.theironfiles.co.uk

References

1. Simon Martin Dyson, Hala Abuateya, Karl Atkin, Lorraine Culley, Sue Elizabeth Dyson and Dave Rowley (2010) Reported school experiences of young people living with sickle cell disorder in England British Educational Research Journal [Research funded by the Economic and Social Research Council, Grant RES-000-23-1486].



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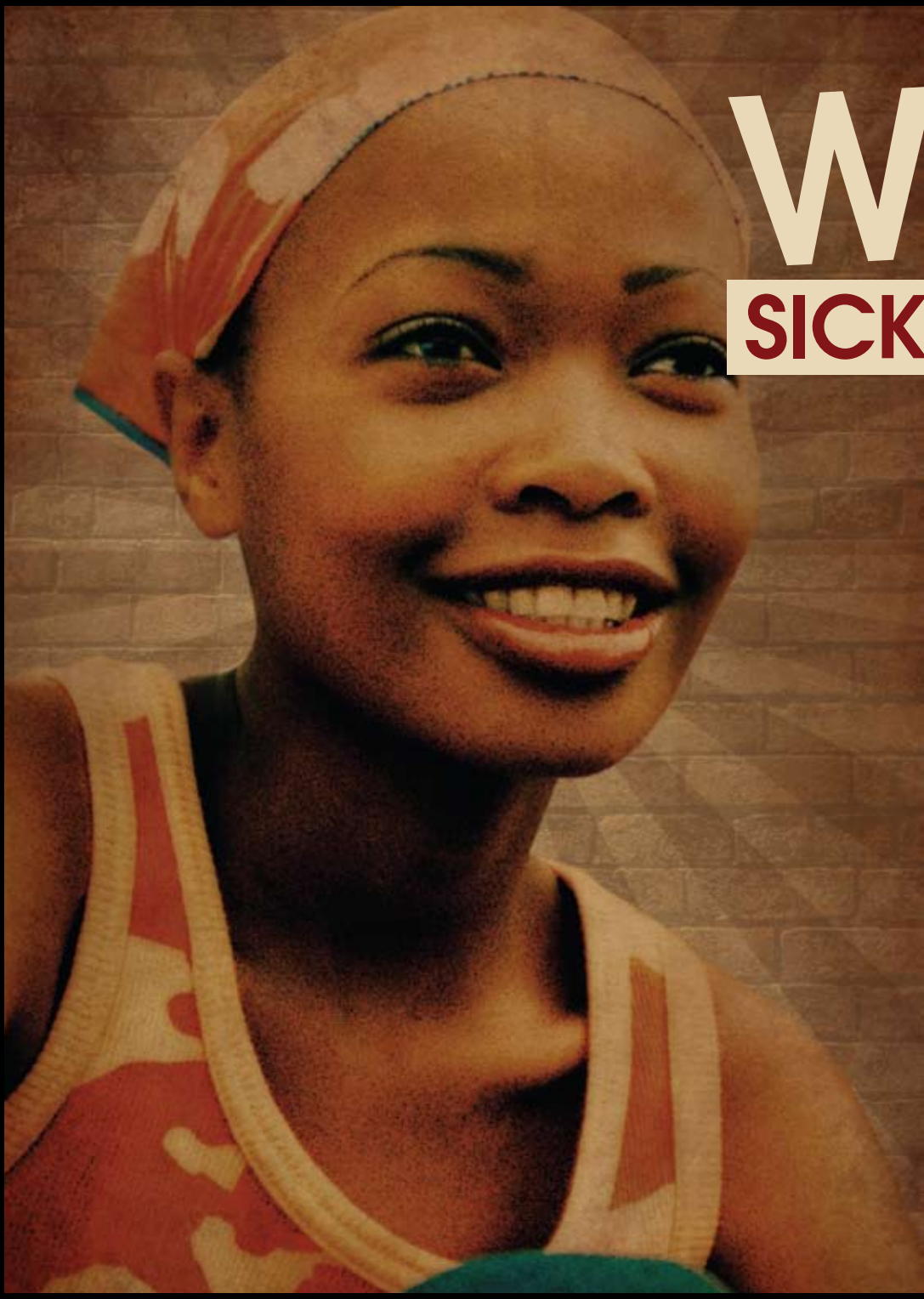
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SICK

WHAT IS SICKLE CELL DISEASE?

Sickle cell disease is a disorder of the blood that may affect every part of the body. It is a disease that is inherited (passed down from parents to child) and is cause by two genes that are not working properly. It cannot be caught.

There are around 15,000 adults and children living with sickle cell disease in Britain at the moment. The most severe form of the disease affects 1 in 50 people of West/Central African descent and 1 in 300-400 of people of African-Caribbean descent.[1]

Sickle cell disease affects the red blood cells. Normally, red blood cells carry oxygen from your lungs to the rest of your body and then carry carbon dioxide back to your lungs for you to breathe out. These red blood cells are round and flexible, shaped a bit like a doughnut, and this allows them to move easily through your arteries and veins to get to every part of your body.

However, in people with sickle cell disease, the shape and texture of the red blood cells can change. They become hard and sticky and are shaped like a farmer's sickle, which is half-moon shaped.

These 'sickle' cells can jam up in the small blood vessels causing blockages and reducing the amount of blood flowing to that area of the body. These blockages can cause excruciatingly painful episodes called sickle cell 'crises'. As well as being extremely painful, these crises can also damage important organs such as the liver, heart and lungs and if they occur in the brain, they can cause strokes. A stroke is what happens when the blood supply to the brain is cut off.

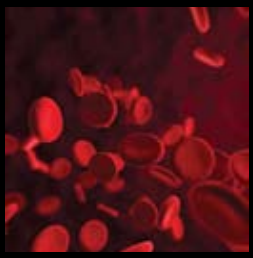
Other complications of sickle cell disease include eye problems, an increased risk of getting infections, chest problems, delayed growth, leg ulcers and anaemia (extreme tiredness caused by having less blood cells than you need).

However, with the right treatment and support, sickle cell disease and its complications can be managed and the risks to your friend reduced.

Know the facts:

- Sickle cell disease is an inherited disease and NOT contagious
- The different types of sickle cell disease are mainly found in people whose families come from Africa, the Caribbean, the Mediterranean, Middle East and Asia although everyone is potentially affected
- Sickle cell disease varies from person to person and not everyone has the same symptoms

HOW DOES SICKLE CELL DISEASE AFFECT YOUR FRIEND?



Normal blood cells

Blood cells of a person with SCD

The symptoms of sickle cell disease can vary from person to person, as can the severity of the disease but the common symptoms are outlined below:

- Anaemia – As your friend's body is not getting all the oxygen it needs, they will feel tired, weak and out of breath
- Hand-foot syndrome – Swollen hands and feet are often the first sign of sickle cell disease in babies. The sickle cells block the small blood vessels in the hands or feet causing them to swell
- Jaundice – This is a yellowing of the skin and eyes caused by the liver not working properly. The liver filters out waste products from the body, and in people with sickle cell it has to work a lot harder to remove the damaged red blood cells
- Frequent infections – The sickle cells can also damage the spleen – an organ that helps the body to develop immunity and fight infections. Failure to develop good immunity will increase their risk of getting an infection. From babyhood people with sickle cell disease are given daily antibiotics to reduce the risk of infections

SICKLE CELL CRISES

Throughout their life, your friend can expect to experience sickle cell crises. Sickle cell crises often occur without warning, or due to a number of reasons such as lack of oxygen, a cold or infection, lack of sufficient water in your friend's body (dehydration) and exposure to extreme temperatures. However, they can occur for no obvious reason. Precautions can be taken to reduce the risk and severity of a sickle cell crisis:

- Drinking plenty of water, and remaining hydrated
- Light exercise
- Eating healthily
- Avoiding extreme temperatures
- Avoiding stress

The severity and frequency of sickle cell crises varies from person to person. Some people experience one or two per year and will be able to control the symptoms using painkillers, whereas others may require hospital treatment.

Symptoms experienced during a sickle cell crisis can include:

- Pain in the bones and joints
- Severe pain in the abdomen (tummy) with the abdomen feeling rigid and inflexible
- Fever
- Difficulty breathing and a stabbing chest pain on breathing
- Weakness on one side of the body and, if the brain is affected, seizures are possible
- Pain in the upper abdomen from the liver and spleen
- Persistent and painful erection of the penis in boys/men

What to do if you think your friend is having a crisis:

- Tell an adult
- Stay with your friend
- Call your friend's parents or dial 999
- Help to make your friend comfortable while you wait for help

TREATMENT

How to manage the condition:

Most of the management of sickle cell disease involves preventing sickle cell crises and treating symptoms and complications if and when they occur.

Minor crises can normally be managed at home using painkillers. However, if the pain is severe or your friend is experiencing the symptoms of a possible dangerous complication, such as chest pain or problems breathing, they may need hospital treatment. Hospital treatment can involve pain relief, blood transfusions, supplemental oxygen and antibiotics.

Sickle-crisis in young people should always be treated under medical supervision due to the risk of infection. So if you believe your friend is having a crisis, it is important that you seek help from an adult immediately.

Some people with sickle cell disease may have to have regular blood transfusions to replace the sickle cells and ensure they have enough red blood cells that are working properly. This may mean that they will occasionally have to miss school to have these transfusions.

WHAT SHOULD I DO NOW?

Your friend's day-to-day life is different from yours; the things you take for granted, they might find difficult so it is important that you show understanding and offer them support when they need it. They don't need wrapping in cotton wool, and probably don't want to be treated any differently to anyone else, but now that you know a bit more about sickle cell disease, it should help you to understand your friend's condition and support them if they need it.