Sickle Cell Unit
UWI, Jamaica.

SICKLE CELL DISEASE IN YOUR TEENS

AN INFORMATION BOOKLET FOR ADOLESCENTS

An open letter to all our adolescents with Sickle Cell Disease

Hey there,

Chances are you've been going to the doctor for your Sickle Cell Disease since you were very little, either with your mum or dad or some other adult. They are the people who, up till now, have known about your disease and what to do when you have problems. You are very lucky that you have had them to look out for you like that.

Now that you are becoming older, it is important that you understand as much as you can about your disease and what **you** can do to help **you** stay healthy.

We hope this book will explain a little about what Sickle Cell Disease is, what you can do to stay well, how to recognise serious problems and what to do if they happen. Most importantly however, we want you to ask us questions – anything you don't understand, we are here to clarify.

We know your teenage years can be challenging but they can be exciting too as you take on new responsibilities and start thinking more about what the future holds. When it comes to things that affect your health... **we are here to help YOU make the right choices for YOU**.

All the best,
The Sickle Cell Team

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The Sickle Cell Unit

Mission Statement

The Sickle Cell Unit is a research institution which seeks to improve the lives of individuals with Sickle Cell Disease through rigorous Biomedical Research, Education, and Clinical Care.

Address: 7 Ring Road, University of the West Indies, Mona, Kingston 7. **Tel:** (876) 927-2471; **Fax:** (876) 927-2984
website: http://www.uwi.edu/tmri/SCU

Hours: Monday - Friday, 8.30 a.m. - 4.30 p.m. (Registration until 12 noon)

The Sickle Cell Unit at the University of the West Indies conducts research to learn more about the condition and treats patients. It has a team of nurses, doctors, lab technologists and support staff dedicated to this purpose. There is a day treatment centre where patients can be admitted during crises and later transferred if they need further care. Patients can come to the clinic for check-ups, for consultations and for treatment. It is important during your teen years, when you begin to take responsibility of your own care, to visit the clinic regularly to monitor your health and progress. Some symptoms may not seem urgent, but may indicate much more serious problems that, if left untreated, could potentially be fatal.

What is Sickle Cell Disease?

Sickle Cell Disease (SCD) is very common in Jamaica, with 1 in 150 people suffering from the said condition. It is an inherited genetic disorder – meaning it is passed down from parents much the same way hair or eye colour is. This particular inherited disease causes a change in the shape of the red blood cells. Red blood cells are usually biconcave in shape and can move easily through blood vessels to supply oxygen to the body. Red cells in SCD change to a 'sickle' shaped red blood cell, which travels less easily through the blood vessels and leads to blockage and subsequent problems in the bones and other vital organs of the body. In addition, the 'sickle' shaped red blood cells carry less oxygen and the cells are destroyed faster than normal red blood cells. This leads to the anaemia (low haemoglobin level) often seen in sickle cell patients.

How long can I live with Sickle Cell Disease?

The average lifespan of persons with SCD is about 55 years but some people die earlier and others live longer. Some early deaths could have been prevented with proper and quick management, if symptoms of more serious problems had not been ignored. Aside from pain, be sure to go see a doctor if you notice paleness, swelling or high or persistent fever. Since sickle cell disease patients are usually anaemic, any worsening paleness (often associated with tiredness) is a sign of either increased destruction or less production of red blood cells. This may be serious enough to require a blood transfusion as a lifesaving intervention. Swelling can occur to bones or joints as a normal part of bone pain in SCD but there are times when the swelling may indicate infection of bone or surrounding tissues. Fever is often a red flag in sickle cell disease and may point to an infection needing immediate attention in order to prevent possible serious consequences.

Signs and Symptoms You Should Not Ignore:

- Increasing paleness
- Fatigue
- Increased yellowing of the eyes
- Fever
- Sudden onset of weakness in arms and legs
- Sudden change in vision

Types of Sickle Cell Disease

There are different types of sickle cell diseases, depending on the specific combination of genes you inherit from your parents – See next page.

The most common are: Sickle Cell Anaemia (SS), Sickle-Hemoglobin C Disease (SC) and Sickle Beta Thalassemia (ST).

Sickle Cell Anaemia: When a child inherits two sickle cell genes, one from each parent, the child has Sickle Cell Anaemia (SS). This type of sickle cell disease is the most common type, and tends to be more severe than others.

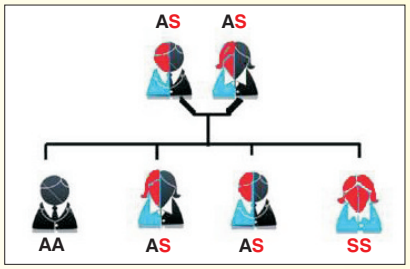
Sickle-Hemoglobin C Disease: Individuals with Sickle-Hemoglobin C Disease (SC) inherit an S gene from one parent and a C gene (which leads to the production of a C type of haemoglobin) from the other parent. Hb SC disease tends to be milder than Hb SS disease, but some people still have severe problems.

Sickle Beta Thalassemia Disease: Individuals with Sickle Beta Thalassemia (Sß-thal or ST) disease inherit an S gene from one parent and a thalassaemia gene from the other parent. The thalassemia gene leads to the production of normal haemoglobin A, but less of it than a normal gene. Sometimes the thalassemia gene leads to the production of NO haemoglobin A. In that case, the child gets $S\beta^{0}$-thalassemia, which is just like Hb SS disease. If some haemoglobin A is produced, but still not normal amounts, the child has Hb $S\beta^{+}$ -thal disease, which is similar to Hb SC disease in severity.

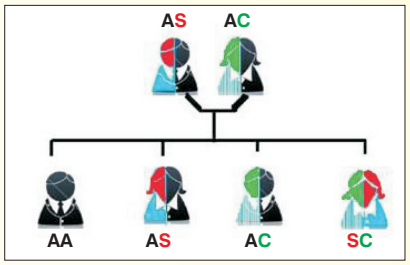
What type of Sickle Cell Disease do you have?

How did you inherit Sickle Cell Disease from your parents?

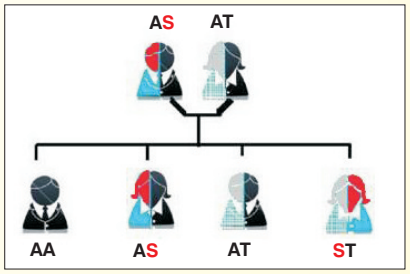
S: Sickle Cell Trait (AS)
Sickle cell trait occurs in 10% of Jamaicans and SS in one in every 300 births. The risk of having a child with SS is 1 in 4 at every pregnancy for parents with sickle cell trait



C: Hb C trait (AC)
Hb C trait occurs in 3.5% of Jamaicans and SC in one in every 500 births. The risk of having a child with SC is 1 in 4 at every pregnancy if one parent has AS and the other has AC trait.



T: Beta-thalassaemia trait (AT)
Thalassaemia trait occurs in 1.5% of Jamaicans and ST (sickle-beta thal) in one in every 3000 births. The risk of having a child with ST is 1 in 4 at every pregnancy if one parent has AS and the other has AT trait.

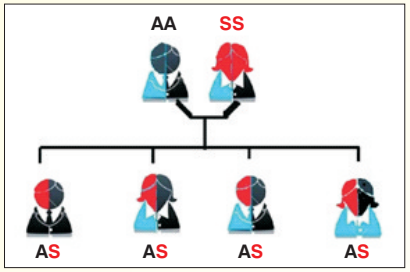


Persons with haemoglobin traits, including Sickle cell trait (**AS**), Haemoglobin C trait (**AC**) or Beta-thalassaemia trait (AT) are perfectly healthy and do not get sick. You will not know they have the trait unless they do a special blood test.

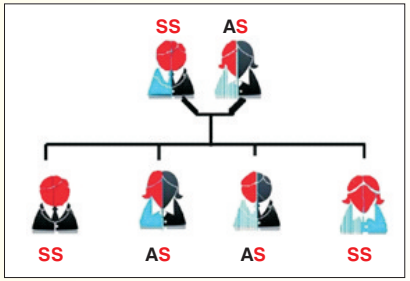
So as you can see there are different types of Sickle Cell Disease. The most common types in the Caribbean are **SS**, **SC**, or **ST** (S-Beta thalassaemia).

When you are ready... Who is the best person to have children with?

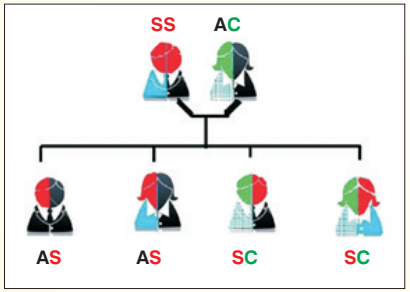
If you have SCD, it is best if your partner has AA. All your children will have sickle cell trait, but none will get sickle cell disease.



If you have SCD and you choose a partner with sickle cell trait, there is a 1 in 2 (50%) chance that your children could have SCD.



If you have SCD and you choose a partner with another trait, such as Hb C trait, there is a 1 in 2 (50%) chance that your children could have a type of SCD.



Remember:

If you have sickle cell disease and you have a child with someone who also has sickle cell disease, **ALL your children with have Sickle Cell Disease.**

What are the symptoms of Sickle Cell Disease?

Having had Sickle Cell Disease from birth, you may have already experienced one or more of the symptoms listed below. This is not an exhaustive list of the complications of the disease, just the most common.

Painful episodes (known as crises)

Some of these painful episodes are brought on by triggers. These include chills, emotional upsets or stress (such as exam stress) or right before a menstrual cycle. You can help prevent the onset of these crises by staying warm and dry and drinking plenty of water. Take pain tablets to alleviate pain, and come to the clinic if the pain is too severe, you notice swelling or if you have a fever or vomiting.

Anaemia:

Because your red cells breakdown a little faster, you may occasionally appear pale or feel tired. If this worsens, you should seek medical attention.

Jaundice (Yellow eyes)

Rapid destruction of red blood cells causes the eyes to become yellow. This is not normally serious, but if you notice your jaundice is worse than usual, you should go see a doctor, especially if you are already otherwise sick.

Shortness of breath

Shortness of breath can indicate Acute Chest Syndrome, one of the leading causes of death among people with Sickle Cell Disease. It is characterized by chest pain, cough, shortness of breath, and/or fever. If you experience any of these symptoms, immediate medical attention is needed as it can be treated very quickly and successfully when detected early.

Ankle sores or ulcers

Leg ulcers are painful and may take a long time to heal. You can prevent them from developing by wearing shoes that fit well and cleaning and covering scratches. If a sore does develop, visit the clinic to get it cleaned and dressed and to get advice on how to help it heal. The sore may need extra treatment if it becomes sloughy.

Sickle Cell Disease in Your Teens

Growing up is tricky enough on its own, but having Sickle Cell Disease can add certain complications. Your teenage years are, in some ways, the riskiest because it is the first time you may be exposed to alcohol, drugs and sex. It is sometimes easy to forget that these activities can have extra adverse effects on you because of your sickle cell disease. This can be dangerous and so, in many ways, you have to be more careful than others.

School

It is important to try, as much as possible, not to let your condition affect your schoolwork. You might miss more school because of your illness. If you have fallen behind on work, don't be afraid to ask teachers for help or get in touch with other students. Sometimes you may need special assistance at school because of your SCD. For example, if suffering from leg ulcers, it can be arranged to have your leg elevated while at school and physical activity limited. Talk to your teachers about your health problems to find an arrangement that allows you to work as best as possible at school.

Puberty

SCD can cause delayed development. What this means is that your body may start to change later than your friends' and classmates'. Teenagers with Sickle Cell Disease can take longer to grow taller and can be smaller than others of the same age. Boys may not notice changes in their voice until later in their teenage years, and may take longer to start growing facial hair. Girls may take longer to start developing breasts and start their periods. The good news is that most people with SCD 'catch up' and there is no significant difference between people with Sickle Cell Disease and non-Sickle Cell Disease individuals by the time they are in their early 20s.

If you are 16 and still have not seen any signs of puberty, visit the clinic to determine if you need to be seen by a specialist.

Alcohol and Drugs

Alcohol is a drug, with dangerous side effects when used in excess. Drinking alcohol in excess dehydrates the body, which affects the blood and can lead to a painful crisis. Extensive alcohol use also causes liver damage. While many teenagers experiment with alcohol during adolescence, for individuals with Sickle Cell Disease, doing so could lead to severe consequences.

Smoking cigarettes is known to be detrimental to your health. However, in Jamaica, adolescents do not associate these risks with smoking marijuana. But, smoking marijuana does carry risks, especially for individuals afflicted with Sickle Cell Disease. Smoking any substance, marijuana or tobacco, is bad for the lungs and smoking may increase the risk of getting acute chest syndrome.

Social Life & Psychological Well-Being

While individuals with Sickle Cell Disease can lead a largely normal life, there are certain precautions that should be taken in daily life with regards to maintaining your health. It is important to exercise and participate in sports if desired, but only within your limits. Stay well hydrated and rest if you feel fatigue.

Certain careers are not suitable for individuals with Sickle Cell Disease, especially those involving a high amount of physical activity. Occupations such as fire-fighters, policemen, athletes or construction workers are inadvisable, and opportunities in industries such as the military or aviation are limited.

The most trying times in your adolescent life can have a severe emotional impact, and may not always have symptoms. If you are feelings overwhelmed by your health problems, confused about sex or development, or are being bullied at school, it is very important to talk about your problems with adults.

Sex

We hear a lot about sex and the various risks it involves, emotional as well as physical – for example, the risk of sexually transmitted diseases and pregnancy. However, Sickle Cell Disease can complicate the matter even further. If you do decide to have sex, you should educate yourself well on the risks involved in sexual activity and the methods of contraception. The risks are not limited to just parenthood, which is a huge responsibility, but also the potential health risks for you and your child.

As a person with Sickle Cell Disease, your child will inherit your genes, including those which cause Sickle Cell Disease. Your partner needs to be tested for the disease as well – if they also have Sickle Cell Disease, your child will be born with the disease. If they have sickle cell trait (AS) or any other gene abnormality causing for example, C trait or thalassemia trait, your child will also have a possibility of being born with sickle cell disease. It is therefore crucial to get your partner tested before becoming pregnant.

(For the girls)

If you are participating in sexual activity, it is important to protect yourself against pregnancy. It is sometimes thought that women with Sickle Cell Disease cannot get pregnant, which is untrue. Even with Sickle Cell Disease, you can get pregnant. There are many contraceptive options available to girls to prevent pregnancy. This is very important as pregnancy in women with Sickle Cell Disease can have serious consequences to both the mother and unborn child.

Condoms: Available at most pharmacies and supermarkets, these are very effective when used properly. Read the instructions on proper condom usage and disposal, and use one every time you have sex.

The Pill: Take one pill a day, at the same time for 3 weeks, and then stop for a week during which you will have your period. Missing pills weakens its effectiveness, so be sure to remember to take one each day. The pills will not become fully effective as you start taking them and so you should also use a condom for the first month of starting them.

The Patch: A small patch worn on the back, buttocks, lower stomach or arm, the patch is replaced on the same day every week for 3 weeks. It is not worn on one week, in which you'll have your period.

The Injection: Called Depo-Provera, it is given every 3 months at a clinic. It tends to stop your periods after 2 or 3 injections have been taken.

Choosing a contraception method with which you are comfortable is a personal decision and involves a period of adjustment. Talk to your doctor about which one you would like to use.

Condoms should still be used even with contraception to protect against the transmission of sexually transmitted diseases.

If you do get pregnant...

Pregnancy requires a lot of extra care, especially if you have Sickle Cell Disease. You should visit the clinic regularly throughout your pregnancy, as women with the disease will need extra care and so will also be referred to a High Risk Antenatal clinic. Pregnant women with Sickle Cell Disease are also more likely to have premature births and smaller babies. Also, painful episodes may worsen or occur more often. There is increased risk of infections during pregnancy including chest, bladder and kidney infections which may also trigger pains. Some women become severely anaemic and require blood transfusion during pregnancy. There is also a known increased risk of death due to pregnancy in Sickle Cell Disease compared to persons without the disease. If you are being treated with hydroxyurea, you should stop the treatment as soon as you find out you are pregnant.

(For the boys)

Sickle cell disease can lead to a condition known as priapism (pronounced pry-ah-piz-im), which is a painful erection of the penis which may last for minutes or hours. If it lasts for 3-4 hours, it becomes a hospital emergency (known as Major Priapism). It is a common problem for boys and men with Sickle Cell Disease, especially likely to occur at night. An acute attack of priapism can be helped by painkillers, hydrating yourself, taking a warm bath or shower, emptying your bladder and gentle exercise (such as walking around the room). It is very important to seek medical help if an erection lasts for an extended period of time as if it is left untreated, it could lead to impotence (meaning an inability to get an erection in the future).

Contraception is also a man's responsibility as well as a woman's. Talk to your partner about birth control, and always use a condom if participating in sexual activity to prevent the spread of sexually transmitted diseases.

Common Myths

Myth	Truth
Only people of African descent have SCD.	Mediterranean people including Greeks and Asians and now all mixed races can have SCD.
People with SCD only live to their 20s	On an average people live into their 50s.
You can catch Leg Ulcers &/or Jaundice from persons with sickle cell disease	You can catch STDs or infections from anyone but you can't catch leg ulcers or jaundice.
Women with SCD are infertile - they can't get pregnant (mule)	Women with SCD are fertile but have a higher risk of problems during pregnancy and childbirth.
If you already have a child with sickle cell disease, the chance that the next one will be affected is much less	Your chance of having a child with SCD is the same with each pregnancy.
You can tell from looking at a person that he/she has sickle cell disease because they look thin.	People with SCD may be thin and/or be jaundiced but sometimes you just can't tell.
You can usually tell by the way they look, that a person has sickle cell trait.	There is NO way to tell by looking. You MUST do a blood test to tell.
In sickle cell disease, white cells eat the red cells.	White cells cannot eat red cells. Red cells are broken down more quickly because of damage from sickling.
Beetroot will make the blood stronger	The anemia in SCD is not caused by iron deficiency anemia and too much iron can sometimes be bad. People with SCD should eat a healthy diet with lots of fresh fruit, vegetable and iron rich foods, but not take iron supplements unless directed by their doctor.

Test Yourself !

1. Sickle Cell Disease is:
   a. Inherited from parents
   b. Caught from another Sickle Cell Carrier
   c. Develops early in childhood

2. When you have the disease, it is important to manage it by:
   a. Staying warm, staying dry, keeping hydrated and getting plenty of rest.
   b. Exercising as much as possible.
   c. Doesn't need management, it doesn't affect everyday life.

3. Sickle Cell Disease affects the shape of:
   a. White blood cells
   b. Red blood cells
   c. The kidneys

4. True or false? There is no treatment for Sickle Cell.

5. Which ones of the following careers are not suitable for individuals with Sickle Cell Disease? : teacher, lawyer, doctor, footballer, banker, policeman, chef, journalist, fire-fighter.

6. Which of the following is NOT a common symptom of Sickle Cell?
   a. Jaundice
   b. Leg ulcers
   c. Bleeding gums

7. True or false: If your girlfriend is on the pill, you do not need to use condoms.

Answers

1. Sickle Cell Disease is:

   Inherited from parents

2. When you have the disease it is important to manage it by:

   Staying warm, staying dry, keeping hydrated and getting plenty of rest.

3. Sickle Cell Disease affects the shape of:

   Red blood cells

4. True or false? There is no treatment for Sickle Cell. False - Drugs like Hydroxyurea can help in cases of severe disease.

5. Which ones of the following careers are not suitable for individuals with Sickle Cell Disease? : teacher, lawyer, **footballer**, doctor, banker, **policeman**, chef, journalist, **fire-fighter**.

6. Which of the following is NOT a common symptom of Sickle Cell?

   Bleeding gums

7. True or **false**: If your girlfriend is on the pill, you do not need to use condoms.

   The pill may prevent pregnancy, but does nothing to stop the spread of sexually transmitted diseases.

Emergency Numbers

Ambulance: Dial 110

Hospital Directory:

- Andrews Memorial Hospital: 926-7401
- Annotto Bay Hospital: 996-2222
- Black River Hospital: 965-2212
- Bustamante Hospital for Children: 968-0300-9
- Cornwall Regional Hospital: 952-5100
- Falmouth Hospital: 954-3250
- Hargreaves Memorial Hospital: 961-1589/ 962-2070
- Hope Institute: 927-2111
- Kingston Public Hospital: 922-0210-9
- Linstead Hospital: 985-2359
- Mandeville Regional Hospital: 962-2067
- May Pen Hospital: 986-6307
- Medical Associates Hospital: 926-1400-1
- Mount Hope Medical Centre: 953-3649
- National Chest Hospital: 927-0000
- Noel Holmes Hospital: 956-2733
- Percy Junior Hospital: 964-2322/2222/4851
- Port Antonio Hospital: 993-2646-8
- Port Maria Hospital: 994-2228/2277
- Princess Margaret Hospital: 982-2304/6
- Savanna-la-mar Hospital: 955-9944/6
- Spanish Town Hospital: 984-3031-5
- St Ann's Bay Hospital: 972-2272-3
- St. Joseph's Hospital: 928-4955
- University Hospital: 927-1620

Suicide Hotline: 1-888-429-5273. A 24-hour hotline to call if suffering from depression for any reason or contemplating suicide

Notes: