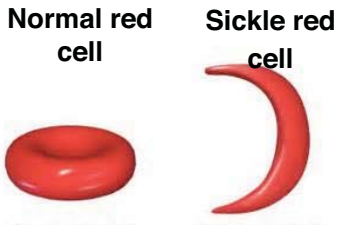
What is Sickle Cell Disease?

Sickle cell disease (SCD) is an hereditary disease of the red blood cells. The main job of red blood cells in the body is to carry oxygen. The protein in the red cell that carries oxygen is called haemoglobin. Persons with SCD inherit abnormal haemoglobin genes from each parent which results in the "sickle change", the production of abnormal red blood cells (**sickle cells**). Normal red cells are soft and round and can travel through the circulation without any problems. **Sickle cells** are hard, sticky and curved in shape (like a banana). They do not live long and are destroyed rapidly in the circulation. Because they are hard and sticky, they may also get stuck and cause blockage of blood vessels. When this happens, some parts of the body will not get enough oxygen. This is what causes many of the problems such as anaemia (weak blood), jaundice (yellow eyes), gallstones, bone pain, pneumonia, leg ulcers (sores), increased susceptibility to infection and strokes that can occur in persons with SCD.

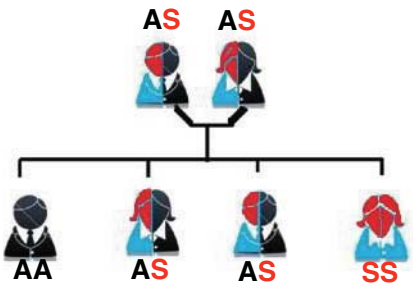


ARE THERE DIFFERENT TYPES OF SCD?

There are many forms of the disease (genotypes). If one inherits the abnormal sickle haemoglobin (Hb S) gene from both parents, the affected person has the most common, as well as one of the most severe forms of the disease (Hb SS disease). If one inherits the sickle gene as well as the gene for another abnormal haemoglobin (eg Hb C) or the gene that does not allow the body to make enough haemoglobin (thalassemia), other forms of SCD may occur such as Hb SC disease and sickle thalassemia (Hb ST). Hb SS disease is often referred to as "full blown" which can lead to the misconception that other forms of the disease are not important. On the contrary, whereas Hb SS disease is a severe form, it is NOT the only severe form of the disease, and even so called MILD forms of the disease can have many problems. In Jamaica, 1 in 150 births are affected by some form of the disease each year.

WHAT IS SICKLE CELL TRAIT?

Persons who have one normal haemglobin (Hb A) gene and one sickle haemoglobin (Hb S) gene are said to be carriers of the disease. They have the sickle cell trait (Hb AS). Other carriers include haemoglobin C trait (Hb AC) and thalassemia trait (Hb AT). Carriers usually have no signs or symptoms and so are often unaware of their status. 1 in 10 persons in Jamaica are carriers of the sickle gene and another 1 in 15 persons carries another gene which puts them at risk of having a child with SCD.



If both parents are carriers of the sickle trait they have a 25% chance of having a child with SCD for EACH pregnancy. *See the diagram above.*

What is Sickle Cell Disease?

COMMON PROBLEMS SEEN IN SICKLE CELL DISEASE

Sickle Cell Disease can affect most if not all of the organs in the body. Some of the common problems include:

- **ANAEMIA** or "weak blood" is due to a lower number of red blood cells in the body. This results in a low blood count.
- **INFECTION** may occur due to a weak immune system and the spleen not working properly. The spleen normally helps to filter germs out of the blood to help prevent infection. Sickle red cells can get trapped in the spleen causing damage and preventing it from working normally. Children are at highest risk of getting serious infections. Special vaccines and antibiotics are given to help prevent infection in children with SCD.
- **PAIN** occurs when sickle red cells get trapped and stuck in the small blood vessels. This affects the circulation of blood and prevents oxygen from flowing to the area where blood cells are trapped. Pain can occur in any part of the body in persons with SCD.
- **ACUTE CHEST SYNDROME** is similar to pneumonia in unaffected persons. However, because it occurs not only from infection, but from other causes such as trapping of sickle red cells in the lung (sequestration) and blockage of small blood vessels in the lung preventing oxygen from getting to the lung, it is called acute chest syndrome.
- **GALLSTONES** occur frequently in persons with SCD because the red blood cells break down much faster than normal red blood cells. The red cells release bilirubin, a substance, which collects in the gall bladder and forms a sludge or gallstones.
- **JAUNDICE** or "yellow eyes" also occurs as a result of increased bilirubin from rapid break down of sickle red cells.
- **STROKE** occurs when the blood does not circulate properly to the brain because of sickle red cells blocking blood vessels in the brain. Children with SCD can get strokes.
- **DECREASED URINE CONCENTRATION** occurs because of small amounts of damage to the kidney. Patients need to drink more fluids and often require more frequent trips to the bathroom as they pass urine more frequntly.
- **PRIAPISM** is a painful, purposeless erection due to trapped sickle cells in the penis. If untreated and persisting for long periods it can lead to erectile dysfunction.
- **LEG ULCERS** or sores often occur around the ankles in persons with SCD. The skin around the ankles gets weak because of poor circulation and can break down easily on its own or from minor trauma such as insect bites.

What is Sickle Cell Disease?

COMMON PROBLEMS SEEN IN SICKLE CELL DISEASE continued

- **SPLENIC SEQUESTRATION** is a sudden enlargement of the spleen due to trapped sickle red cells, with a drop in the blood count. It is most common in young children and can cause death. Parents / guardians can be taught how to check for an enlarged spleen so that they can seek medical attention for their child quickly.
- **SICKLE RETINOPATHY** (eye disease) occurs from damage to the back of the eyes (retina). It can lead to blindness. Persons with SCD should have their eyes checked each year, from age 12 years, to look for early signs of eye disease.

IMPORTANT FACTS ABOUT SCD

1. Sickle cell disease (SCD) is a genetic disease.
   - Persons with disease inherit the genes from BOTH parents.
   - It is NOT an infection. Problems of the disease such as leg ulcers (sores) and jaundice cannot be "caught".
   - The risk is the same (1 in 4) for EACH pregnancy when both parents have a trait.
2. 15 % of Jamaicans carry genes that put them at risk for having a child with SCD.
   - 1 in 10 persons are carriers of the sickle gene.
   - A simple blood test can tell you whether you are at risk.
3. Babies can be tested for SCD from immediately after birth.
   - 1 in 150 babies born in Jamaica will have a form of SCD.
   - Children are at risk of dying early from complications of the disease.
   - Early diagnosis of disease allows for simple treatments to be started which can help prevent death in early childhood.
4. SCD is NOT a "black person" disease.
   - Whereas it is more common in persons of African descent, the disease also occurs in other races – Caucasians, Asians and people from the Mediterranean regions.
5. Persons with SCD do not all die young.
   - The lifespan of a person living with a severe form of SCD is, on average, ten years less than persons without disease. Simply put, the average age at which people in Jamaica with a severe form of SCD die is approximately 55 years.
   - Early diagnosis, being informed about their disease and good health care will help patients and their family to learn how to deal with their disease so that they can prevent complications or respond to them quickly when they occur.

What is Sickle Cell Disease?

6. Children with SCD can learn like other children and SHOULD be encouraged to go to school as much as possible.
   - They may have to miss school often because of sickness. Parents and teachers should work together to ensure that work is sent home and where possible extra help should be given to these children to allow them to catch up with their school work. If you notice a change in your child's ability at school - speak to your doctor as it may be a sign of a mild stroke.
   - They should be encouraged to participate in all school activities (including sports) to their **PERSONAL LIMIT**.
7. Adults with SCD often have difficulty maintaining a job, especially if they get sick often.
   - Employers should be informed about the disease, problems their employees may have and how best that they can support them.
   - Patients and employers need to work together to ensure that they remain productive members of the workforce.
8. Children with SCD can get strokes.
   - A simple ultrasound test is now available at the Sickle Cell Unit, which can help identify children at higher risk of having stroke.
   - There are treatments that can be started in children to lower their risk of having a stroke.
9. Women with SCD are NOT infertile.
   - They do, however, have an increased risk of complications during pregnancy and delivery.

IMPORTANT HEALTH TIPS FOR PERSONS WITH SCD

Here are some tips to help you / your relative deal with sickle cell disease to prevent complications and know how to respond to them when they occur:

1. Get informed about sickle cell disease.
2. Keep appointments with your doctor for regular check-ups.
3. Ensure that you receive all available immunizations to prevent serious infection.
4. Eat a balanced diet, with lots of fruits and vegetables.
5. Drink plenty of fluids, especially water.
6. Avoid extreme cold, wet or heat.
7. Keep pain medication at home. Pain often starts during the night or early morning.
8. Avoid overexertion.
9. Exercise to your PERSONAL LIMIT.
10. Avoid stressful situations.

Preventing Infection

Patients with sickle cell disease (SCD), especially Hb SS and Hb SB0 thalassemia are at increased risk of life-threatening infections. This is because the immune system in patients with SCD does not function normally. This allows bacteria (germs) to grow in the blood and can lead to serious infections especially in young children. Serious infections can cause disability and death.

The table below shows some of the bacteria (germs) that cause major problems in persons with sickle cell disease and how to prevent these problems.

STREPTOCOCCUS PNEUMONIAE (Pneumococcus)	This bacterium (germ) causes serious infections including infection of the blood and of the lining of the brain. Patients with Hb SS & Hb Sβ0 thalassemia are given penicillin – ***Penadur*** (injection every 28 days) or ***Penicillin V*** (medicine to take by mouth twice daily) – from ***age 4 months to at least age 4 years*** to prevent infection with this germ. If taken regularly, penicillin is extremely effective in reducing the risk of infection and death. If you are allergic to penicillin – don't worry – there are other options. ***Pneumococcal vaccines also help prevent infections*.** **PCV** – given at age 2, 4, and 6 months with a booster at age 15 months **Pneumovax** – given at age 4 years with a single life-time booster after 5 years.
HAEMOPHILUS INFLUENZAE TYPE B (HIB)	This is another bacterium (germ) that can cause serious infection and death in sickle cell patients. A ***vaccine*** is available to help prevent infection with HIB. It is given at age 2, 4, and 6 months with a booster at age 15 months.
SALMONELLA	This is a bacterium (germ) that causes bone and blood infection in sickle cell patients. Most infections occur as a result of eating infected food like chicken and eggs, although you can get it in other ways. To prevent salmonella infection: • Cook all meats (especially chicken) thoroughly. • No raw or undercooked eggs (e.g. soft-boiled or "scald" eggs). • Wash all utensils and dishes properly after using with raw meats. • Practice proper hand washing before handling / eating food and after using the bathroom. • No unusual pets (e.g. turtles and lizards).

Preventing Infection

HEPATITIS B	This is a virus (another type of germ) that can cause long-term illness that leads to liver disease (cirrhosis), liver cancer and death. It is spread through contact with the blood and body fluids of an infected person (e.g. having unprotected sex, sharing needles and blood transfusions). ***Hepatitis B vaccine*** can prevent infection. You will need 3 doses of the vaccine for full immunization.

Signs and symptoms of infection include:

- Fever greater than 38°C/101°F
- Shortness of breath or difficulty breathing
- Pale colour of the tongue or lips
- Markedly increased jaundice (yellowing of the eyes)
- Weakness or unusual tiredness
- Unusual pain crises

If you note **any** signs or symptoms of infection you should go to your nearest doctor or hospital as soon as possible.

The earlier treatment is given the better, however, even with treatment permanent disability and death can occur.

INFECTIONS IN SICKLE CELL DISEASE CAN CAUSE SERIOUS PROBLEMS AND CAN OFTEN BE PREVENTED.

1. Ensure all immunisations are up to date
2. Practice good hygiene
3. Cook all foods thoroughly
4. BE ALERT for signs / symptoms

SEEK MEDICAL ATTENTION EARLY!

Ask your doctor for further information.

Nutrition in SCD

There are several lines of evidence that suggest that nutrition is important in SCD. For example the chronic increased breakdown and production of red blood cells in sickle cell disease increases the rate at which body protein is broken down and rebuilt (protein turnover). Additionally, because the body has to work harder to ensure all the tissues get enough oxygen, the overall metabolic rate is increased. As a result, the body requirements for carbohydrates, protein and fats are higher in SCD compared to the average person. The consequences of the increase nutritional requirements are varied but include delays in growth and maturation, and thinness (a body mass index or BMI <18.5 kg/m^2), which is more common in adults with SCD. It is generally accepted that persons who are thin tend to have more complications when they are ill.

In addition to the global increased requirements for energy and protein there appears to be an increase in the specific demands for various nutrients. Some of these are:

- Amino acids –arginine, citrulline, and praline.
- Minerals & trace elements – magnesium and zinc.
- Fatty acids - n3-fatty acids, short chain fatty acids.
- Vitamins - vitamin E and folic acid.

Another abnormality that has been described in SCD and which may respond to nutritional intervention is increased oxidative stress. This occurs when oxidative molecules which are by-products of normal metabolism are produced in excessive amounts. Oxidative molecules are neutralized by antioxidant molecules. Failure to neutralize oxidative molecules results in damage to cells which can lead to worsening of the illness. There is a large body of literature which has reported that the antioxidant capacity of red blood cells of individuals with SCD is low. The importance of this observation is that, oxidative damage of the red cell membrane of persons with SCD alters the ability of red cells to bend .This contributes to the formation of sickle shaped cells that can cause blockage of blood vessels in different organs in the body. Therefore eating foods that increase the antioxidant capacity of individuals may be beneficial in SCD.

Individuals with SCD are encouraged to choose a diet with a wide variety of foods from all food groups each day. Each day include items from **starches** (eg. yam, potato, breadfruit, rice), **protein** (eg. red meat, fish, chicken), **fruits** (eg. mango, oranges, banana, tomato), **vegetables** (eg. carrot, lettuce), **dairy products** (eg. milk, cheese) and **fats** (eg. fish oil, margarine, butter).

Often times during a painful episode the appetite will be poor. If this occurs one can take protein-energy supplements.

Management of Bedwetting

Unlike other children, many children with sickle cell disease (SCD) have difficulty concentrating their urine. As a result of this, during the daytime they will pass urine more often and at night they may have to wake up to pass urine. Waking up when the bladder is full is very difficult even in kids who do not have sickle cell disease. Bedwetting is therefore more common in children with sickle cell disease.

Children who are still wetting the bed beyond the expected age for becoming dry at night, usually by age 6, may benefit from the following measures:

1. **Avoid** the following drinks from about 6pm: milk, tea, chocolate or cocoa, cola sodas, alcohol. These drinks increase the amount of urine we make.
2. The child should **empty the bladder** just before bed-time. **Double voiding** is always useful – that is encouraging your child to try to pass urine again 5 minutes after he/she last voided.
3. The last adult to retire at night should **awaken** the child and see that the bladder is again emptied.
4. If the toilet is far from the child's bed, have a **potty** available at the bedside, especially for younger children. A night light is also useful to help older children find their way to the toilet.
5. Get an **alarm clock** and set it to wake the child in the early hours of the morning to pass urine again.
6. Get a personal calendar for the child and give **reward** points for dry beds. Gradually increase the number of dry nights that are required to obtain the reward. Start with realistic goals – 1 dry bed/week and increase as there is improvement.
7. Use plastic to protect the mattress and get your child to help to change the bed sheets.
8. Quarrelling, embarrassing or humiliating your child will only make the problem worse. Encouraging your child to wake-up and being patient will help them to achieve dry nights.

Praise works!

9. **Bladder exercises** sometimes help. During daytime, the child should hold his/her urine for as long as possible and then empty the bladder. The stream of urine should then be briefly halted during the passage of urine and the process then continued to the point of emptying the bladder completely.

Your child may never be able to sleep though the night without passing urine… they must therefore learn to wake up to pass urine in a potty or the toilet.

Managing Pain at Home

PREVENTION

- Keep warm especially when the weather is cool.
- Avoid getting wet in the rain and do not keep on wet clothes.
- Drink a lot of water – get a water bottle to take with you **at all times**.
- **EVERYONE IS DIFFERENT** - know what brings on your pain and avoid those situations.

Examples of medications you can use at <u>home</u>

Drug	Frequency	Examples of brand names
Paracetamol	(4-6 hourly)	Panadol, Tylenol, Cetamol, Tempra, Calpol, Tylex
Aspirin (over 12 yrs)	(6 hourly)	Codis, Disprin, Excedrin
Paracetamol + codeine	(4-6 hourly)	Panadeine, Cetadine , Panadeine F*
Ibuprofen	(6 hourly)	Advil, Brufen, Motrin
Diclofenac*	(8 hourly)	Voltaren, Cataflam
Codeine*	(4-6 hourly)	
Tramadol*	(8 hourly)	Tramal

***Prescription required**

Mild - Moderate pain: **A) Paracetamol every 4 hours**
B) Paracetamol + Ibuprofen <u>or</u> diclofenac OR
C) Paracetamol and codeine (e.g. Panadeine)

Moderate - Severe pain: **A) Panadeine + Ibuprofen <u>or</u> diclofenac OR**
B) Paracetamol + Ibuprofen <u>or</u> diclofenac + codeine <u>or</u> tramadol

Many of the above medications are available over the counter – i.e. no prescription is necessary

Important points:

1. Start pain medication early at the first sign of pain
2. Start with medication appropriate for the amount of pain you are having and <u>add</u> medication as necessary if the pain is not getting better or if it is getting worse.
3. Rest, increase oral fluids and eat regularly – most pain medications should not be taken on an empty stomach, however paracetamol works best when taken on an empty stomach.
4. Take medication **regularly** for 24 hours minimum even if you are feeling better. This will prevent the pain from returning.
5. As the pain improves, stop the strongest medication you are taking but continue to take the others; if the pain does not return, discontinue the next strongest and so on until you are off all your medication.
6. Most pain can be managed at home if you start your medication early and take it regularly.

SEEK MEDICAL ATTENTION: if your pain is not settling after 48 hrs, if it is getting worse despite appropriate medication or if there are other concerns such as high fever,shortness of breath, vomiting or weakness.

ALWAYS WALK WITH YOUR WATER BOTTLE & SOMETHING TO KEEP YOU WARM!

HOME PAIN PLAN

If your pain is:	Use these Medicines	Key Points
LEVEL 1: Mild Pain Pain Scale: 1-3	_____ _____ _____	• Give medicine by the clock – **DO NOT WAIT FOR THE PAIN TO RETURN** • Once **PAIN-FREE** for at least 24 hours, **STEP DOWN** medicine • **STEP DOWN** medicines by one (1) level every 24 hours if pain does not return • If pain worsens, **STEP UP** medicine for 24 - 48 hours then try again to **STEP DOWN** • Remember to **drink extra** fluids and to **keep warm** • Visit the doctor or the clinic if your pain is not settling, is getting worse or if you develop other symptoms that you are worried about.
LEVEL 2: Moderate Pain Pain Scale: 4-6	_____ _____ _____	
LEVEL 3: Severe Pain Pain Scale: 7-10	_____ _____ _____ _____	

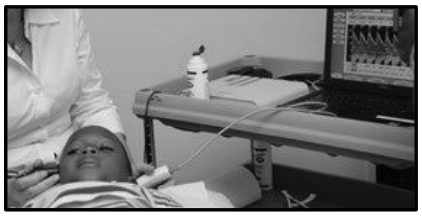
Management Plan for: _____

Please review this plan yearly with your doctor.

Stroke and TCD Screening

What is a stroke?

Strokes occur because of a blood circulation problem to the brain. If the blood flow is decreased, parts of the brain do not get the oxygen necessary for the brain to function. A stroke may result in problems such as weakness in an arm or leg, difficulty talking or understanding what others are saying, memory problems or other losses of brain function. These problems may be temporary or permanent. If you see these signs in yourself or your child see your doctor urgently. Early treatment can limit the disability that can occur after a stroke.



What is transcranial doppler (TCD)?

TCD is a test that uses ultrasound (similar to the ultrasound used during pregnancy) to detect areas of increased flow in the blood vessels of the brain. The TCD test is painless and harmless. Your child will need to lie on a bed and we will put a small amount of gel on a flat probe and place it on each side of your child's head (just above and in front of the ear). This is where we will best be able to hear the blood flow in the vessels we are testing. The test usually takes about 30-40 mins but may take longer in younger children. You may stay with your child during the test. Your child must be well (no recent illnesses) and must stay awake during the test.

What will the TCD result tell me?

TCD measures how fast the blood is flowing through the blood vessels supplying the brain. When blood vessels are narrowed due to sickle cell damage, the blood flows more quickly through those blood vessels. When blood vessels are narrow, it is easier for them to become blocked and so the child is at a higher risk for having a stroke.

What happens after the test?

It may take a few days to read and interpret the test results. You will be seen again to discuss the test results. If the test is normal, nothing needs to be done, although your child will be retested once a year until the age of 16 years. If the test results are positive (findings of fast blood flow, indicating possible vessel narrowing) or questionable (either because the test was difficult to do, or because the results are only slightly positive), another TCD screen will be scheduled. Other tests such as a MRA (magnetic resonance angiography) - another painless imaging test, may be advised at that time.

It is important to understand that TCD is a test that tells us if a child may be at risk for having a stroke *in the future*. If your child displays any of the symptoms of having a stroke (e.g. sudden weakness in an arm or leg, difficulty speaking, seeing or understanding), you must see a doctor *immediately*. If in doubt, go to the nearest Accident and Emergency so that other tests can be done to determine if your child is having a stroke.

What if the results show that my child is at higher risk for having a stroke in the future?

If the test is abnormal 2-times in a row, then your doctor will discuss treatment options with you. In many countries, blood transfusions are given every 4-6 weeks to prevent a stroke from happening. A study has shown that regular blood transfusions, in children who have abnormal TCD tests, decrease the risk of getting a stroke in the future. ***Transfusions, once started, must be continued for life.***

In many countries, including Jamaica, it is very difficult to arrange for blood transfusions every month for children with SCD. If your child's TCD is not normal, and you live in a place where blood transfusions cannot be done regularly, your doctor will discuss using a medication called hydroxyurea (HU) instead. There is some evidence in the medical literature that HU can decrease TCD levels. This should reduce the risk of having a stroke. A study in Jamaica has also shown that, in children who have had a stroke, HU can decrease the risk of having another stroke.

Leg Ulcers

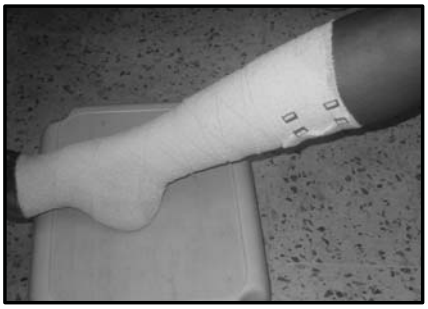
In Jamaica, leg ulcers occur in up to 30% of persons with sickle cell disease.

Prevention:

- Protect your ankles from biting insects/scratches/trauma especially in childhood.
- Wear proper fitting shoes and sandals.
- **Massage and moisturize** the skin around the ankles daily.
- Daily exercise of feet.
- Elevate feet when sitting.
- Clean and dress cuts and scrapes as soon as they occur.

If you get an ulcer:

1. See your doctor as soon as possible if you think you are developing an ulcer.
2. If your ulcer is sloughy, your doctor will advise you on what you can do to get it looking red and healthy again so that it can heal. This may involve using grated green pawpaw (papaya) wrapped in gauze on the ulcer to get rid of the slough.
3. Twice daily cleaning with diluted hydrogen peroxide is ideal (1 part hydrogen peroxide to 6 parts water) when the ulcers are sloughy, but once they are clean – clean only once daily.
4. Daily application of gauze dressing soaked in saline or antiseptic solution like potassium permanganate solution – (it should be a very light pink colour when made up)
5. Keep dressings wet when the ulcer base is red and healthy looking. If they dry out, soak the dressing with clean water or saline before removal. Adaptic® dressings are very good for this as they do not stick to the ulcer and may be tried.



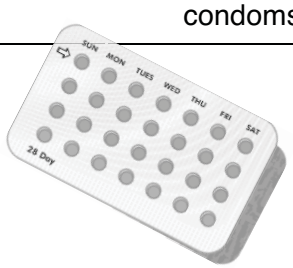
6. Elastic supportive bandage or compression stockings should always be used during the daytime – the picture shows you how you should wrap your ankle.
7. Keep your leg up as much as possible during the day to prevent swelling of the ankle.
8. Twice daily massage of the skin surrounding the ulcer (you may use cocoa butter, olive oil or any other emollient available) and do ankle exercises (roll your ankle in circles 12 times clockwise and 12 times anti-clockwise). This improves circulation in your foot and may help to decrease the swelling.
9. Taking zinc and multivitamins may help ulcers heal more quickly.

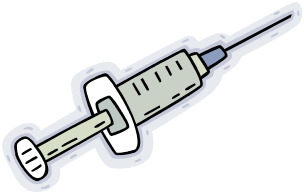
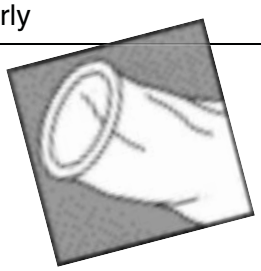
AFTER YOUR ULCER HAS HEALED:

- **YOU MUST BE EVEN MORE CAREFUL TO PROTECT YOUR ANKLES.**
- **CONTINUE TO WEAR COMPRESSION STOCKINGS AS THIS MAY PREVENT SWELLING OF THE ANKLE THAT LEADS TO RECURRENCE OF THE ULCERS.**

Female Issues in SCD

Things to consider.	• You may notice that you start seeing periods later than other friends or family without sickle cell disease - this is normal for persons with sickle cell disease. You can usually expect to see your periods about two years after you start developing breasts. If you are not having periods by the time you are 16 years old then see your doctor. • If you are having sex but do not want to get pregnant then you have **MANY** options for family planning!! – see below. • If you are planning to get pregnant - ask your partner to get tested for sickle cell disease so that you know the chances of passing the disease on to your child. • If you become pregnant, you should see a doctor as soon as you miss a period! Your sickle cell disease may become worse while you are pregnant - so your pregnancy should be watched VERY CLOSELY by a team of doctors who know about sickle cell disease.
What are my family planning options?	**YOU CAN USE ANY TYPE OF FAMILY PLANNING METHOD THAT OTHER PATIENTS CAN USE!!** • An injection every 12 weeks (3 months) called depo provera • Any type of family planning pills (called “the Pill”): these you take one (1) pill every day for 21 days, then take a break for 7 days when you will have a period and then repeat this again with a new pack of pills • Family planning patches: you wear one (1) patch on your arm or another part of your body and change it every week for three (3) weeks, then take a break for a week when you will have a period and then repeat this again with a new pack of patches • A very small coil can be put into your womb by a specialist doctor to prevent you from getting pregnant, your specialist will tell you when to change it • You or your partner can use condoms: female or male condoms work well when used properly



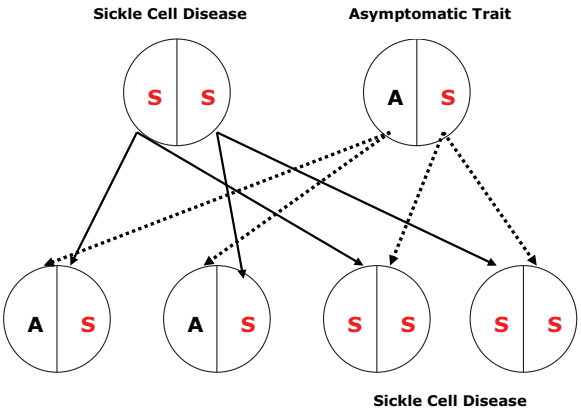



Female Issues in SCD

What if I become pregnant?	• You may have problems with high blood pressure while you are pregnant. • You may have worse or more frequent pain. • You may have more serious infections. • Your blood count may fall very low - you may need to get blood (this is called a blood transfusion). • You may get chest infections (like pneumonia) more easily while you are pregnant. **WHEN YOU BECOME PREGNANT...** • Register early at a high-risk antenatal clinic or with a high-risk specialist obstetrician. • Take folic acid and iron supplements right through your pregnancy. • You will be told to see your specialist doctors more often than other patients who do not have sickle cell disease. • You may need to spend more time in hospital when you have your baby if you have any problems at the birth or shortly after that.

Remember:…..

- If you are using hydroxyurea (HU) to help with your sickle cell disease then you will need to stop using it six (6) months before you get pregnant.
- Try to plan every pregnancy to protect your health and the health of your baby.
- Because you have sickle cell disease, you **will** pass a haemoglobin gene that can cause sickle cell disease to your baby. Before you get pregnant, it is best to check if your partner has AA type blood. That way your baby will not get sickle cell disease.
- If your partner has the sickle cell trait. There is a 50% chance your baby will have sickle cell disease



A Patient's Guide to Hydroxyurea

HOW DOES HYDROXYUREA WORK?

Haemoglobin (Hb) is the oxygen carrying protein in red blood cells. Most persons have Hb A in their red blood cells but persons with sickle cell disease (SCD) have inherited a gene from their parents that causes their body to make a different type of haemoglobin called Hb S. As blood travels through the body, haemoglobin in red blood cells picks up oxygen from the lungs and carries it to all other parts of the body. In SCD blood vessels may become blocked depriving different parts of the body of oxygen. This causes pain and over time can also damage organs and affect their proper functioning.

Babies born with SCD do not show symptoms of the disease right away. This is because when babies are born, they have a special type of haemoglobin called Hb F. This type of haemoglobin protects the baby from the complications of SCD. Over the first 6 months of life, as Hb F levels decrease and Hb S levels increase, the infant eventually starts having symptoms related to SCD.

Hydroxyurea (HU) is a drug known to increase the amount of Hb F in the red blood cells. Persons with SCD who take HU show an increase in Hb F levels in their red blood cells over time and this may protect them from some of the complications of the disease. There are likely to be other ways in which HU helps prevent sickle cell problems, but these are not clearly understood.

WHAT ARE THE POSSIBLE BENEFITS OF HU THERAPY?

Hydroxyurea is the only drug approved for the treatment of sickle cell disease. **HU has been shown to reduce the frequency of many of the complications such as painful crises, acute chest syndrome, the need for transfusions and hospitalization by at least half with minimal toxicity.**

WHAT ARE THE RISKS?

There are certain risks and discomforts that may accompany HU therapy. HU can reduce the white cell count and decrease the body's ability to fight infections. It can occasionally cause upset stomach, vomiting, skin rashes and hair loss. Experience tells us though that these bad effects are **uncommon** and happen in **less than 1 %** of all patients taking the drug. **They also usually go away quickly when the drug is stopped.**

WHAT ARE THE POSSIBLE SIDE EFFECTS?

Frequency	Side effect
Common (> 20%of persons)	**Early**: Decreased white blood cell count – increased risk of infections Decreased platelets – increased risk of bleeding
Occasional (5–20% of persons)	**Immediate**: stomach upset **Early**: mouth ulcers, anaemia, hair loss
Rare (<5% persons)	**Early**: skin rashes, dizziness, headaches, confusion, hallucinations, seizures **Delayed**: skin colour and nail changes, bleeding, fever & infections **Late**: possible cancer risk, possible birth defects

A Patient's Guide to Hydroxyurea

IS HYDROXYUREA RIGHT FOR YOU OR YOUR CHILD?

YES if you have Hb SS or HbSβ^0 thalassemia **and one or more of the following:**

- Recurrent painful crises
- Recurrent acute chest syndrome
- Severe symptomatic anaemia
- In children following a stroke if transfusion programs are not offered

NO if you are pregnant, wish to have children in the near future or have active liver disease.

BEFORE HYDROXYUREA IS STARTED…

There are a number of things that have to be done before hydroxyurea is started:

- Baseline blood and urine tests will be done.
- All post pubertal females will be expected to do a pregnancy test and if not already on contraception, one will be recommended for them. Contraception will also be discussed with male patients and their partners.
- Your immunization status will be reviewed and updated (this helps to reduce the risk of infections).
- Once hydroxyurea is started you will also be expected to take folic acid daily.

HU is usually started at a low dose and slowly increased over time if the patient remains well. Additionally, simple blood tests can sometimes inform the doctor if a potential side effect is developing before the patient has any symptoms at all. It is for these reasons that people who take HU need to be seen regularly for a check up and for blood tests.

AFTER HYDROXYUREA IS STARTED…

- Initially, you will be seen every 4 weeks and blood tests will be done at those visits.
- Keep your appointments – though problems might prevent you from attending clinic from time to time, persons who regularly miss appointments may not be able to continue taking hydroxyurea.
- Report possible problems.
- Tell your doctor immediately if you think you may be pregnant.

Live a healthy life style: eat well, keep well hydrated, exercise, and do not drink alcohol or smoke

Priapism

Priapism is a ***persistent, painful, penile erection***. It occurs mainly in adolescents and young men BUT can occur at any age, and has been seen in boys as young as age 2 years.

In males with sickle cell disease, there are two main types of priapism:

Stuttering priapism

These are episodes that can last up to 2-3 hours, but that resolve without any medical treatment. They frequently occur at night and may or may not be associated with sexual activity. Often though, there is no obvious trigger. Stuttering priapism can lead to major priapism so it is very important to advise your doctor if this is happening to you.

Major priapism

When a penile erection lasts longer than **4 hours**, it is called a major priapism. An episode of major priapism is an emergency. Because there are often delays getting to a hospital, males with priapism should make their way to an Emergency Room if an episode of priapism has not resolved within 2 hours.

If priapism continues beyond 4 hours, there is an increased risk of permanant damage to the penile structure, which may lead to erectile dysfunction such as impotence.

What can you do if you have an acute attack of priapism?

- Take pain medication e.g. paracetamol, ibuprofen, codeine
- Drink extra fluids
- Take a WARM bath / shower
- Empty your bladder
- Do some gentle exercise (walking around the room, sit-ups)

IF SIMPLE MEASURES AT HOME ARE NOT HELPING AND PRIAPISM IS LASTING LONGER THAN 2 HOURS, GO TO YOUR NEAREST HOSPITAL IMMEDIATELY.

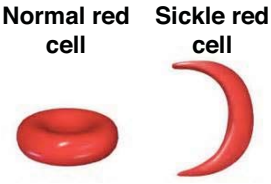
REMEMBER:

- Prolonged episodes can lead to erectile dysfunction, such as impotence, in the future.
- Prevention of episodes can be difficult; however, if you or your child is having stuttering episodes, let your doctor know as soon as possible. There are medications that may be able to control these episodes and so prevent a major attack.

A Teacher's Guide to Sickle Cell Disease

What is Sickle Cell Disease (SCD)?

SCD is an inherited blood condition which affects about 1:150 children born in Jamaica. It affects the red, oxygen-carrying protein, haemoglobin (Hb), in the red cells. Most people have Hb A but when you have SCD most of your haemoglobin is Hb S. This causes the red cell to change shape.



(See "What is Sickle Cell Disease?" for more information.)

What are the symptoms?

- Rapid destruction of red cells causes anaemia (weak blood). Your student may tire more easily especially during PE classes. This increased break down of red cells also causes jaundice or yellowing of the eyes.
- The sickle shaped cells may also block blood vessels in the body which can lead to pain, breathing difficulties and strokes in children.

Can a child with SCD participate in school activities?

Absolutely! They should be encouraged to participate in all school activities **BUT** to their personal limit. They should be allowed to stop if they start having pains or feel short of breath. Additionally, they should be encouraged to drink extra fluids as dehydration can cause sickle cell problems.

Getting cold and wet can occasionally bring on pains, so swimming may be a problem in some children but everyone is different. In general, they should not play in water for long periods, and should dry off quickly and change into warm dry clothes ASAP.

There are two exceptions where children should not be allowed to participate in sports:

- Avascular necrosis (AVN) of the hip – this is damage to the hip joint from poor blood flow. Children with AVN should not weight bear and should use crutches at **all** times when moving about, to help their hip to heal.
- Leg ulcers – these sores about the ankle are a common problem, especially in adolescents and they may take a very long time to heal. They are not infectious. Keeping the foot elevated is one of the most important ways to promote healing.

Is there anything you can do?

1. Watch out for teasing. Children can be unkind to other children who are different. Children with SCD tend to be smaller than average, have yellow eyes (jaundice), may use crutches or may have sores on their leg. Protect them from this by explaining the disease to other children.
2. Children with SCD are encouraged to drink a lot of water and they do not concentrate their urine well. You should therefore allow more frequent bathroom breaks.
3. Children with SCD miss more school than average as they may have routine clinic or doctor's appointment or because they are unwell at home or have been admitted to hospital. To make up for these absences, they should be given encouragement, extra help and remedial help. Although there are many people with SCD who have achieved academically, SCD can affect the ability to learn quickly in some children. Falling behind or increasing difficulty with learning may be a sign that the child has had a mild stroke. You should discuss these changes with the parents so they can talk to their doctors. You should also refer for educational assessment, where available, if required. Patience and guidance are essential if the child is to achieve his/her true potential.
4. Look out for signs of new medical problems. If the child looks paler than usual or has a high fever – seek medical help immediately. If they complain of pain, allow them to lie down, give them extra fluids to drink and a pain killer like paracetamol.

You can help prevent or minimise many of the problems of this disease so that the child can grow up to be a productive member of society.