Basic facts about Sickle Cell Anaemia and Pregnancy

A factsheet for NGO staff and non-medical programme people

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What is Sickle cell anaemia?

- Sickle cell anaemia is a genetic inherited blood disorder where the haemoglobin is not normal.
- Haemoglobin (Hb) is a protein in red blood cells that carries oxygen to the tissues of the body.

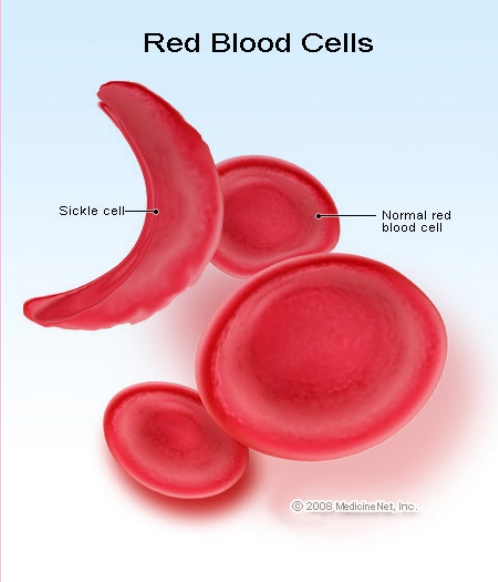
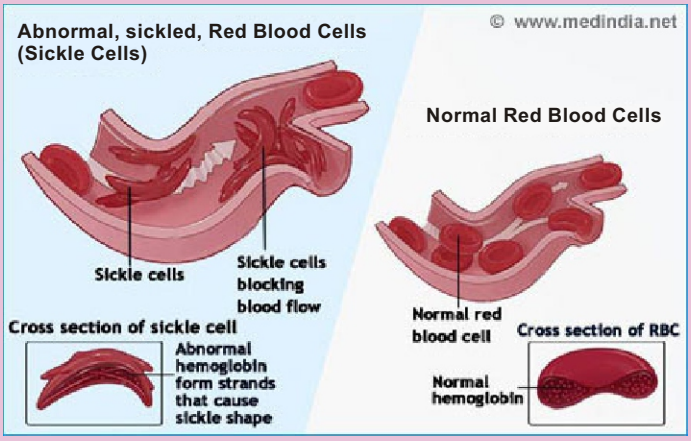


Figure 1: Shape of sickle cells compared to normal red blood cells

- Normal red blood cells (RBCs) are smooth, round, and flexible, like the letter "O," so they can move through the vessels in our bodies easily.
- RBCs with the abnormal haemoglobin are stiff and sticky, and form into the shape of a sickle, or the letter "C", especially when they give up their oxygen to the tissues.
- Normal adult haemoglobin takes oxygen from the lungs to tissues throughout the body and gives it up there. Sickle haemoglobin can also transport oxygen. However, once the oxygen is released, sickle haemoglobin tends to line-up into rigid rods that alter the shape of the red blood cell.



- These sickle cells tend to bunch up and cannot easily move through the blood vessels and cause a blockage. This is more likely to happen when the person has lost a lot of fluids due to sweating, vomiting or does not have enough intake of fluids, during labour etc.

Figure 2: Sickled cells in blood vessels
http://www.veria.com/healing/sickle-cell-anemia-managing-a-life-long-disease

- This blockage leads to the damage of the supplied organ and pain associated with this condition.
- RBCs with sickled Haemoglobin only live for about 15 days, while cells with normal Haemoglobin can live up to 120 days.
- Due to the decreased number of RBCs circulating in the body, a person with sickle cell disease (SCD) is chronically anaemic.

What causes sickle cells to be created?

- Sickle cell disease is inherited.
- It is the result of a genetic mutation which is thought to have originated in areas of the world where falciparum malaria was common, since people with sickle trait are somewhat protected against the deadly falciparum malaria.
- Sickle cell disease primarily affects those of African descent but is also found in some parts of India.

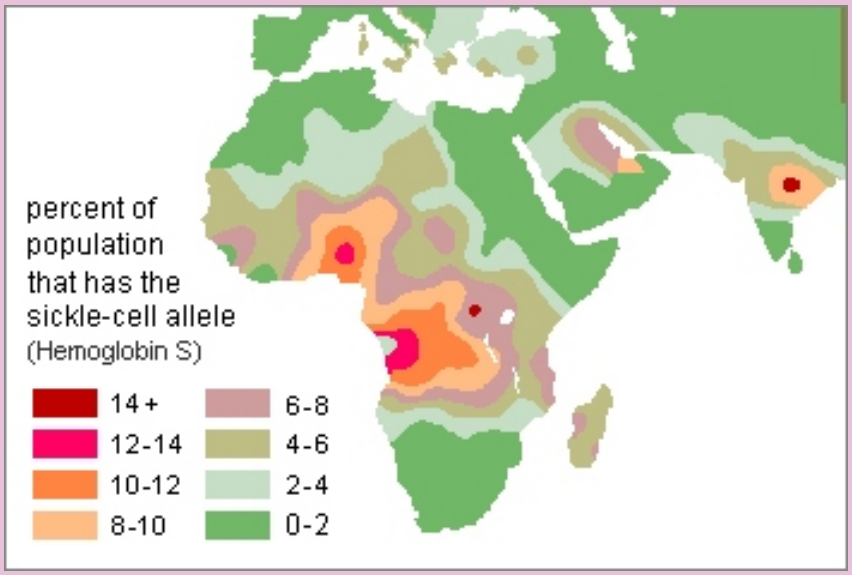


Figure 3: World map showing distribution of sickle cell disease
http://stritch.luc.edu/sickle_cell/content/research-agenda-sickle-cell-disease-africa-unprecedented-opportunities-unsolved-challenges

How many women in India have this condition?

The prevalence of sickle-cell anaemia is high in Central and Southern India, among tribals and in people of some other social groups such as Sahu, Teli, Manikpuri, Vasava, Choudhary, Gamit and Dodhiya[1]. Tribals in the states of Madhya Pradesh, Gujarat, Maharashtra, Chattisgarh, Orissa, Tamil Nadu and Kerala witness a high incidence of the disease. The proportion is as high as 25 percent[1] in some part of the country.

[1] Sickle Cell Anaemia. Kasturba Hospital, Sewa Rural. Jhagadia. Bharuch.

What is Sickle cell trait?

A baby will be born with sickle cell disease only if two sickle cell genes are inherited - one from the mother and one from the father. Persons who have only one sickle cell gene are healthy and said to be carriers of the disease. They may also be described as having sickle cell trait.

What is the risk with Sickle cell trait?

If both parents have the trait then every child they have has a one in four, or 25 percent, chance of being born with sickle cell disease. This means that there is a three of four, or 75 percent, chance for another child to not have sickle cell disease. There is also a 50 percent chance that a child will be born with sickle cell trait, like the parent

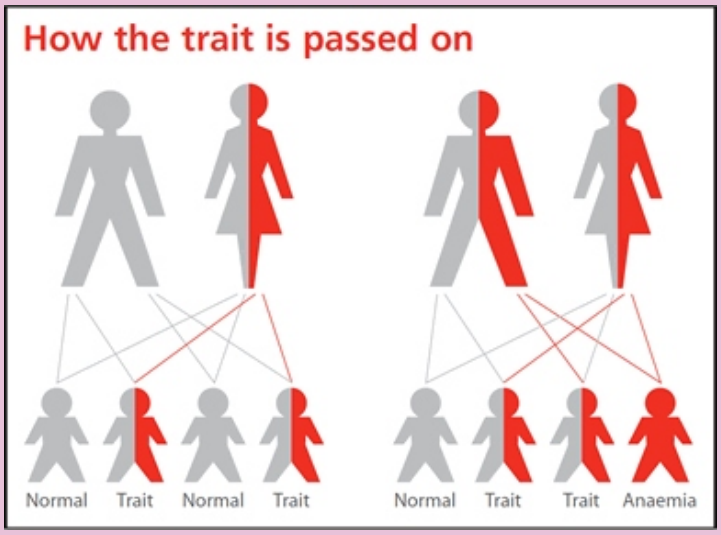


Figure 4: Possible ways of inheritance of Sickle Cell Trait and Disease
http://www.cdc.gov/ncbddd/sicklecell/documents/scd-factsheet_scd--pregnancy.pdf

How does one find out if one has it?

- A simple blood test, done at any time during a person's lifespan, can detect whether he or she has sickle haemoglobin. However, early diagnosis is very important.
- In some countries there is mandatory testing for sickle cell anaemia as part of their new-born screening programs.
- If the test shows some sickle haemoglobin, a second blood test is done to confirm the diagnosis.
- Doctors also can diagnose sickle cell disease before birth using a sample of tissue taken from the placenta or by amniocentesis.

Why is sickle cell disease a problem?

Sickle cells have a shorter life span than normally shaped red blood cells. This results in chronic anaemia characterized by low levels of haemoglobin and decreased numbers of red blood cells.

Sickle cells are also less flexible and stickier than normal red blood cells, and can become trapped in small blood vessels preventing blood flow. This compromises the delivery of oxygen, which can result in painful episodes and damage to associated tissues and organs.

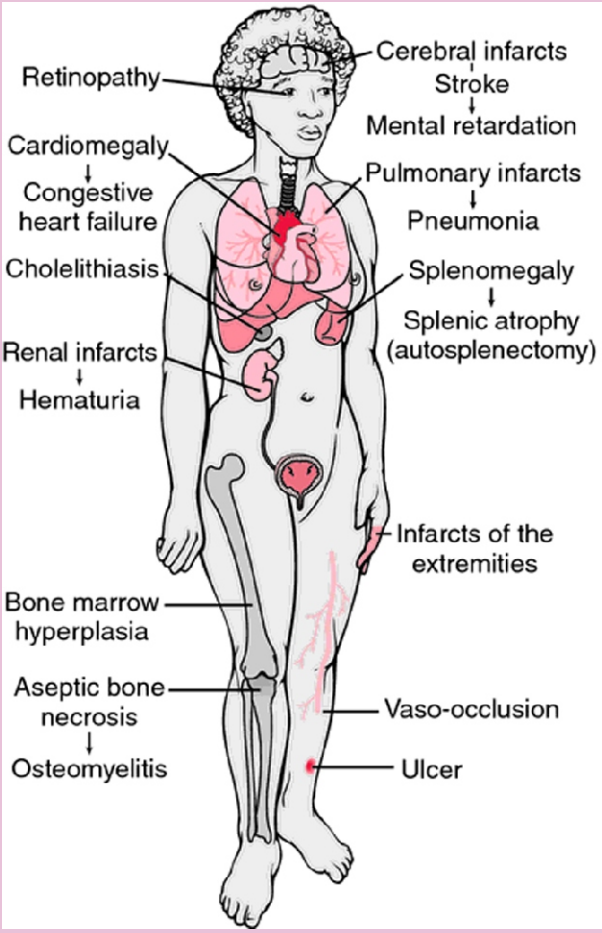


Figure 5: Organs affected by sickle cell disease
http://medical-dictionary.thefreedictionary.com/sickle+cell+disease

What conditions promote the sickling (distortion) of the red blood cells in sickle cell anaemia?

Sickling of the red blood cells in patients with sickle cell anaemia results in cells of abnormal shape and diminished flexibility. The sickling is promoted by conditions which are associated with low oxygen levels, increased acidity, or low volume of the blood. These conditions can occur as a result of injury to the body's tissues, dehydration, or anaesthesia.

Certain organs are predisposed to lower oxygen levels or acidity, such as when blood moves slowly through the spleen, liver, or kidney. Also, organs with particularly high metabolism rates (such as the brain, muscles, and the placenta in a pregnant woman with sickle cell anaemia) promote sickling by extracting more oxygen from the blood. These conditions make these organs susceptible to injury from sickle cell anaemia.

How does sickle cell disease affect pregnancy?

The risks for pregnancy depend on whether the mother has sickle cell disease or sickle cell trait. Generally, women with sickle cell trait are not at increased risk for problems, however, they may experience frequent urinary tract infections.[2]

It is also important to remember that, unlike sickle cell disease, a woman with sickle cell trait can have iron deficient anaemia while pregnant and may need iron supplementation for this reason.

[2] ww.jcdr.in/articles/PDF/1834/45%20-%203466.(A).pdf

The ability of the blood cells to carry oxygen is especially important in pregnancy. The sickling and anaemia may result in lower amounts of oxygen going to the foetus and slowed foetal growth. Because sickling affects so many organs and body systems, women with the disease are more likely to have complications in pregnancy.

What can be the complications for a pregnant woman?

Complications and increased risks for the mother may include, but are not limited to, the following:

- Infection, including urinary tract (especially kidney) and lungs
- Gallbladder problems including gallstones
- Heart enlargement and heart failure from anaemia
- There is an increased risk of sickle cell crisis[3] and Acute Chest Syndrome (ACS)[4] in the intra-partum period and of painful crisis with protracted labour of more than 12 hours, but the latter is often secondary to dehydration.

Complications and increased risks for the foetus may include, but are not limited to, the following:

- Miscarriage
- Intrauterine growth restriction (poor foetal growth)
- Preterm birth (before 37 weeks of pregnancy)
- Low birth weight (less than 2.5 kg)
- Stillbirth and new-born death

[3] **Sickle Cell Crisis:** Acute onset of signs and symptoms which can be fatal if untreated. The patient can present with unbearable joint pain, pain in abdomen, jaundice, pallor, fever with chills, chest pain and breathlessness, convulsions, stroke, unconsciousness, splenomegaly and shock.

[4] Acute Chest Syndrome: Acute onset with symptoms such as cough, chest pain, fever and breathlessness

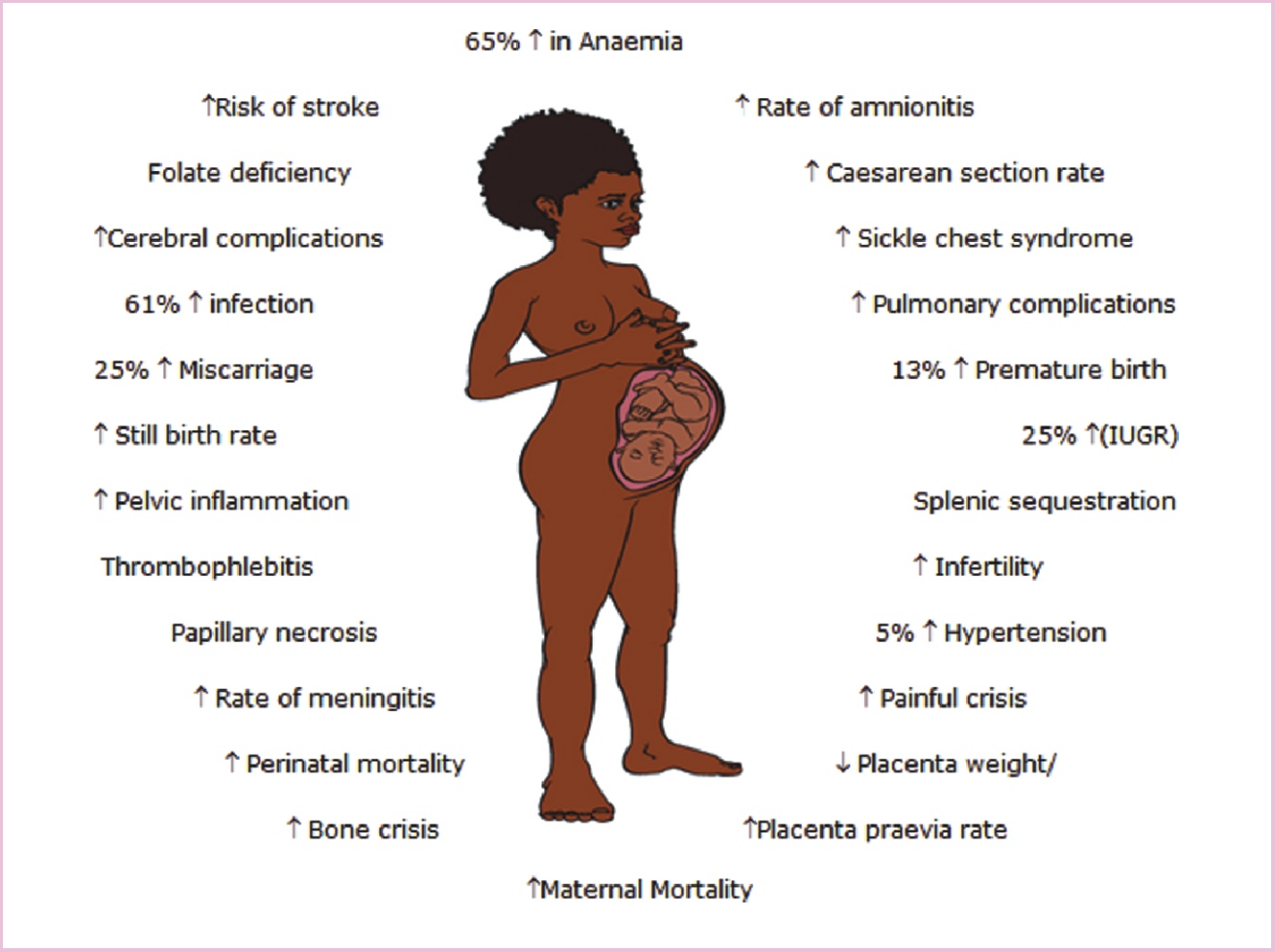


Figure 6: Effect of sickle cell disease on pregnancy
Ref http://medical-dictionary.thefreedictionary.com/sickle+cell+anemia

How should they be managed?

Royal College of Obstetrician and Gynaecologists, UK, Greentop Guidelines http://www.rcog.org.uk/womens-health/clinical-guidance/sickle-cell-disease-pregnancy-management-green-top-61

Women and men with sickle cell trait should be encouraged to have the testing done for their partner determined before they plan a pregnancy.

Antenatal Care:

Early and regular antenatal care is important for pregnant women with sickle cell disease. Blood vessel blockage or painful crises are more common in pregnancy. All women with

sickle cell anaemia should be managed in a tertiary health care (medical college / civil hospitals) with a multi- disciplinary team. While it is true that some women with sickle cell disease have a very turbulent antenatal or intranatal course, a large majority have uncomplicated pregnancies.

1. General pregnancy care should include a healthy diet, prenatal vitamins and folic acid supplements (B vitamin). The woman should drink 10-15 glasses of water every day to prevent dehydration.
2. Excessive vomiting can also lead to sickle cell crisis (sudden increase in sickling caused due to dehydration leading to pain in joints and organs) and should be managed urgently. Aspirin and ibuprofen should be avoided for pain after 30 weeks of pregnancy.
3. Severe anemia is more common in pregnancy. Some women may benefit from blood transfusions. These may be needed several times during the pregnancy to help increase the blood's ability to carry oxygen and decrease the number of sickled cells. Attempt should be to keep the haemoglobin over 9 grams percent.
4. It is important for women who receive blood transfusions to be screened for antibodies that may have been transferred in the blood and that may affect her foetus. The most common antibodies are to the blood factor Rh.

Other management guidelines:

- Hepatitis B vaccination is recommended and the woman's immune status should be determined before she gets pregnant.
- It is recommended strongly that women with sickle cell disease should sleep inside a bednet to prevent malaria. If these women get fever, they should be checked for malaria on priority basis. Any infection should be treated immediately.
- Folic acid (5 mg) should be given once daily both before and throughout pregnancy.
- Routine iron supplementation should be avoided unless the pregnant lady is iron deficient
- Blood pressure and urinalysis should be performed at each consultation, and midstream urine for culture performed monthly.
- Prophylactic blood transfusion is recommended in pregnant women with sickle cell disease who are at high risk of developing complications or crisis.
- Ultrasound screening for assessment of foetal growth restriction and foetal distress in the third trimester should be recommended.

Postpartum care

- In pregnant women where the baby is at high risk of sickle cell disease (i.e. the partner is a carrier or affected), early testing for sickle cell disease should be offered. Capillary samples should be sent to laboratories where there is experience in the routine analysis of sickle cell disease in new-born samples. This will usually be at a regional centre.
- Maternal oxygen saturation should be maintained above 94 percent and adequate hydration should be maintained based on fluid balance until discharge.
- The same level of care and vigilance should be maintained as has been described for antenatal care, since acute crisis and other complications of sickle cell disease remain a risk in the puerperium.

Contraception methods

- Progestogen-containing contraceptives such as the progesterone only pill, injectable contraceptives, and the levenorgestrel intrauterine system are safe and effective in sickle cell disease.
- Estrogen-containing contraceptives should be used as second-line agents.
- Barrier methods are as safe and effective in women with sickle cell disease as in the general population.

What is the outlook for patients with sickle cell anaemia?

The life expectancy of persons with sickle cell anaemia is mildly reduced. Some patients, however, can remain without symptoms for years, while others do not survive infancy or early childhood. Nevertheless, with optimal management patients can now survive beyond the fourth decade.

Causes of death include bacterial infection (the most common cause), stroke or bleeding into the brain, and kidney, heart, or liver failure. The risk of bacterial infections does diminish after three years of age. Nevertheless, bacterial infections are the most common cause of death at any age. Therefore, any signs of infection in a person with sickle cell anaemia must be reviewed with a doctor to prevent damage and save lives.[5]

[5] Medically reviewed by David Hoffman, MD; American Board of Internal Medicine with subspecialties in Oncology and Haematology, Bunn, HF. Pathogenesis and treatment of sickle cell disease. N Engl J Med 1997; 337:762. Harrison's Principles of Internal Medicine, McGraw-Hill, edited by Eugene Braunwald, et. al., 2001.

State level programmes in India

States such as Gujarat, Orissa, and Tamil Nadu have programmes to manage the condition. The aim of these programmes is to ensure that there is no child birth with Sickle Disease by 2020.

http://www.tihf.in/scdcenter.pdf

Some key strategies include:

1. Prevention of spread of sickle cell disease by reducing sickle cell births through screening and genetic counselling.
2. Identification of sickle-cell anaemia in new-born & infants as early as possible.
3. Provision of early treatment to sickle cell patients.
4. Counselling to patients and relatives so that they understand the scope of the problem and participate in the management of the disease.
5. Creation of awareness about the disease among the entire community.

References

RCOG Greentop Guidelines :
http://www.rcog.org.uk/womens-health/clinical-guidance/sickle-cell-disease-pregnancy-management-green-top-61

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