Government of India

Ministry of Tribal Affairs & Ministry of Health & Family Welfare

Counseling Modules on Sickle Cell Disease

Towards
SICKLE CELL FREE INDIA

सत्यमव जयते

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Introduction

Towards Elimination of Sickle Cell Anaemia in India by 2047

The Budget 2023 contained an announcement which reads as **"A Mission to eliminate Sickle Cell Anaemia by 2047 will be launched. It entails awareness creation, universal screening of 7 crore people in the age group of 0-40 years in affected tribal areas, and counselling through collaborative efforts of central ministries and state governments".** The lead Ministry in this regard is the Ministry of Health and Family Welfare (MoHFW) with the supporting Ministry being the Ministry of Tribal Affairs (MoTA). There are other stakeholders identified by the MoHFW in this Mission, including the State Governments.

Constitution of Committee by MoTA:

Following the post budget Webinar on the announcement held on 27.02.2023 and in order to formulate a comprehensive and effective strategy, this Ministry has constituted a Committee to devise guidelines for generating awareness about the disease, its management and prevention of future births with the disease as well as for counseling of patients, care-givers and other stakeholders when an individual is diagnosed with the trait/disease.

Since among communities, there may be certain beliefs, traditional customs, myths, practices related to health and disease, there is need of dedicated efforts to spread scientific awareness about how the disease is transmitted and the manner in which the transmission can be controlled.

These two modules on awareness and counseling are prepared with the aim to provide awareness to the general population as well to the patients, caregivers, school teachers and the health workers. The modules have been prepared in the form of PPT so that it is easier for every one to understand the sickle cell manifestation. In the modules one may find duplication of the messages as these messages have to be conveyed to all the stakeholders. The Awareness module is primarily intended to reach the general public, especially the tribal population, the medical fraternity, the influencers at the local level – the ASHA, the Anganwadi worker, the teacher, the youth, the traditional headmen and healers, the Gram Panchayats and so on. The Counseling module is intended for a smaller target group of those who are directly affected by the disease and are responsible as primary or secondary caregivers such as patients, their parents and other caregivers in the family, school teachers, co-workers and health workers.

The Ministry takes this opportunity to thank the Chairperson and Members of the Committee as well as other experts for having drafted the module within a very short period of intense deliberations. The Ministry also acknowledges the support given by the MoHFW in the vetting, finalization and translation of these documents. It is sincerely hoped that these documents would go a long way in achieving the Mission's objective of elimination of Sickle Cell Anaemia by 2047 in India.

Table of Contents

Module Part - I

Overview of Counseling for Sickle Cell Disease

What is Sickle Cell Anemia?

- Sickle cell anaemia is a genetic condition where the haemoglobin molecule inside red blood cells is abnormal.
- Both men and women have equal risk of getting this disease.
- The red blood cells carrying sickle-haemoglobin become sticky and deformed (C shaped) and get destroyed resulting in anaemia
- Spectrum of Sickle Cell : Carrier AS

  Sickle Cell disease

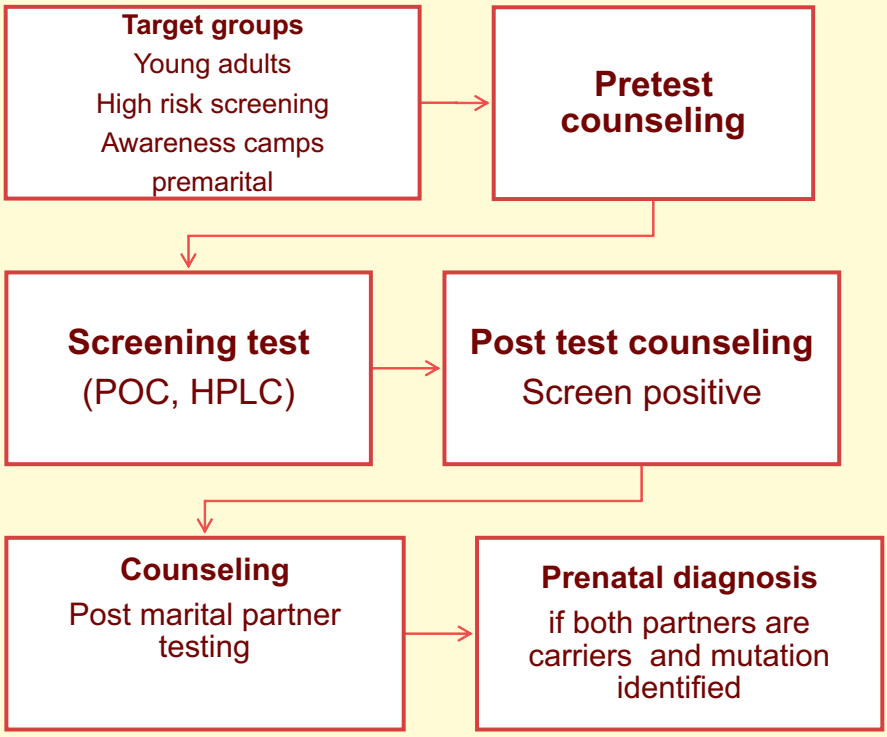
  Sickle Cell Anemia

- This is a chronic disease with need for life long visits
- Sickle cell anaemia occurs in children whose parents are sickle cell carriers
- Since the prevalence of sickle cell carrier state is high in India, it is important to be aware of this condition
- Awareness can improve screening and in reducing the burden of this disease
- Being aware of your sickle cell state is very important before planning a child
- Screening is therefore advisable for

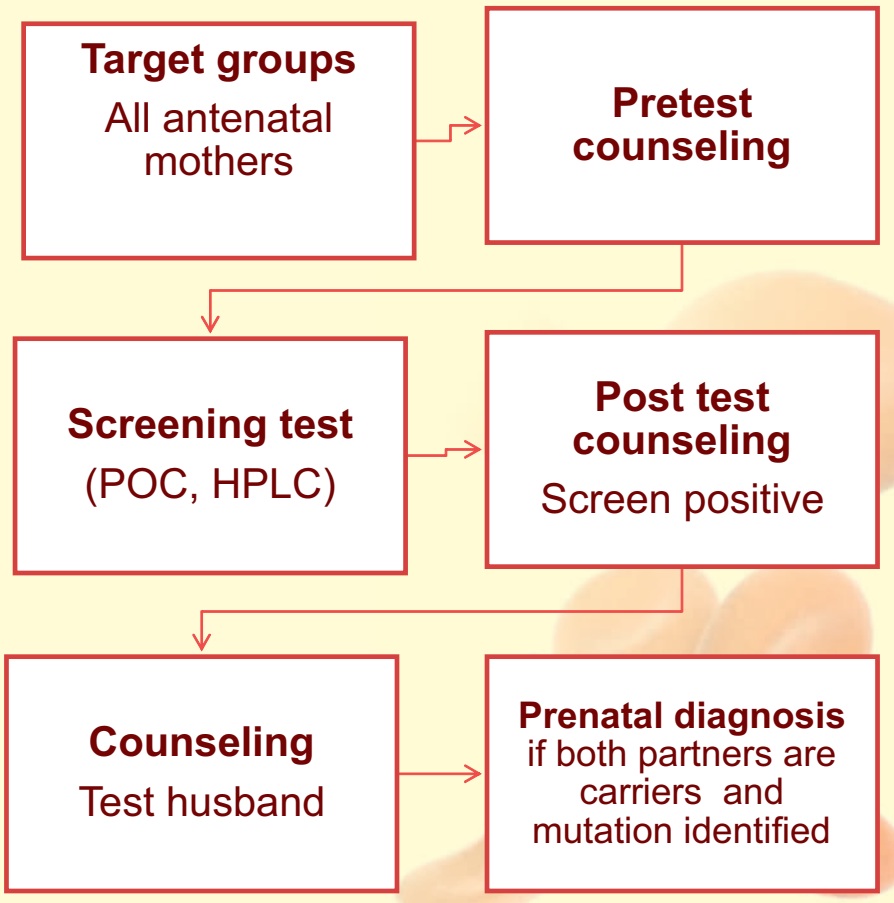
  1. All pregnant women during the first 3 months of pregnancy
  2. All school and college going children in high prevalence areas
  3. Community screening in high prevalence areas (all persons between 0-40 years including newborns)

What is Sickle Cell Anemia?

Population Screening



Prenatal Screening

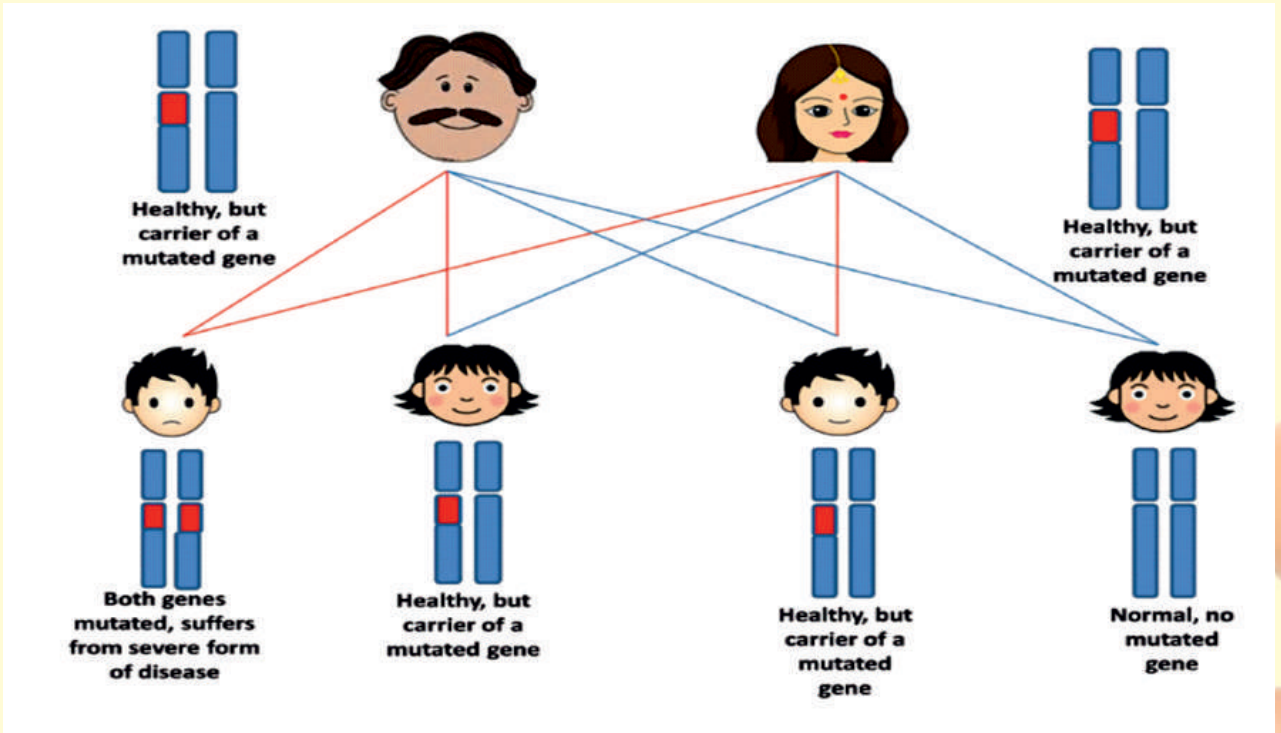


Module Part II

Community screening for Sickle Cell status

After screening counselling for positive patients AS, SS

- Children suffering from sickle cell trait / carrier have one gene of the sickle cell from one of their parent. These children won't have complications and symptoms in general. But they act as a carrier.
- Sickle cell disease can have complications which varies from person to person, and life-long treatment is required.
- The treatment of sickle cell disease is available at primary health centres or community health centres
- It is a genetic change in the blood forming genes
- This is passed from parents to their children
- The affected children can be either male or female
- In every pregnancy there is a 25% risk of having an affected child, if both partners are carriers.



Module Part - III

Screening for
Sickle Cell Carrier status

Pre-test counseling

Antenatal screening for detecting sickle cell carrier state

Why do we screen for sickle cell carrier status in antenatal period?

- The carrier rate of sickle cell disease is high in India. Carriers are not symptomatic. So unless you check your carrier status by HPLC or electrophoresis, you may be completely unaware of this. Hence, always check your carrier status, before you plan on starting a family.
- If you have been detected as a carrier (HbAS), then check the carrier status of your spouse and meet a doctor for further counseling.
- With correct knowledge you can prevent having children with sickle cell disease in your family.

Pretest counseling

- Prenatal testing is available to help you and your partner know whether your baby will have sickle cell disease or not.
- The prenatal test is advised if both of the parent are sickle cell carriers or one of the parent has sickle cell anaemia (HbSS) and the other is a carrier.
- You can prevent the birth of a child with a severe haemoglobin problem like sickle cell disease if you decide that is what you want.
- Prenatal testing is done at 3-4 months of pregnancy at selected centres.

How is the test for detecting sickle cell carrier state done?

- It is done by few simple blood tests
  - Complete blood counts
  - HPLC
- Both are done by drawing 2-3 ml of blood
- The results are usually ready within 2-3 days

एक जांच, स्वस्थ परिवार के नाम

Pre test counseling – Sickle cell carrier state

- Both men and women have equal risk of getting this disease.
- It is a genetic change in the blood forming genes
- This is passed from parents to their children
- The affected children can be either male or female
- In every pregnancy there is a 25% risk of having an affected child if both partners are carriers.

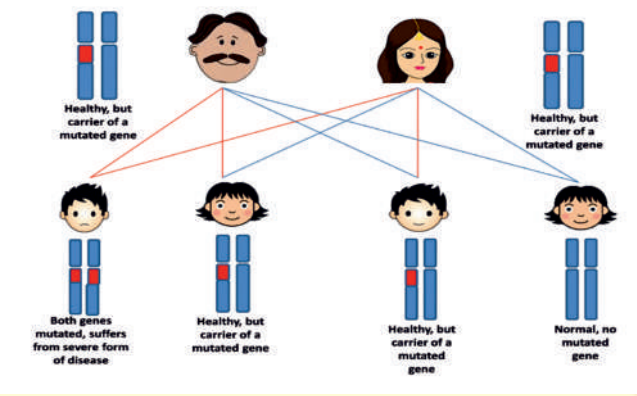
What happens if you are tested to be a sickle carrier?

- If you are tested to be a sickle cell carrier, there is no need to panic
- Sickle carriers live a normal life
- If you are planning a child, then test your spouse immediately
- If you are not, it is still preferable to check the status of your spouse so that you are prepared for this later

What happens if you both are tested to be sickle carriers?

- If both you and your spouse have been tested to be a sickle or thalassemia carrier, then immediately visit a center which has the provision for prenatal testing
- There is a 25% chance of having a baby with sickle cell disease
- Timely prenatal diagnosis with CVS or Amniocentesis will help in diagnosing the disease in the baby
- Do prenatal testing by 12-15th week of pregnancy

Post-test antenatal counselling where mothers and spouses are positive for Sickle carrier state (AS)



- If both pregnant women and her spouse have been tested to be a sickle carrier, then immediately visit a center which has the provision for prenatal testing.
- There is a 25% chance of having a baby with sickle cell disease.
- Timely prenatal diagnosis with CVS or Amniocentesis will help in diagnosing the disease in the baby.
- Do prenatal testing by 12-15th week of pregnancy.
- What Are The Risks With This Result? both sickle carriers) If two sickle cell carriers get married- then they 25% chance of having a child with sickle cell disease

What is the test done to detect the Sickle cell status of the baby?

- If both you and your spouse are sickle cell carriers and if you are pregnant at present, the status of the baby should be known before 20 weeks of pregnancy
- The tests done that can be done for this purpose are
  1. Chorionic Villus Sampling
  2. Amniocentesis
  3. Percutaneous Cord blood sampling
- The tests are done between 10-18 weeks of pregnancy
- A needle is inserted into the uterus and sample is taken from the respective areas
- Results are available within 3-4 weeks

Will this test be harmful for my baby?

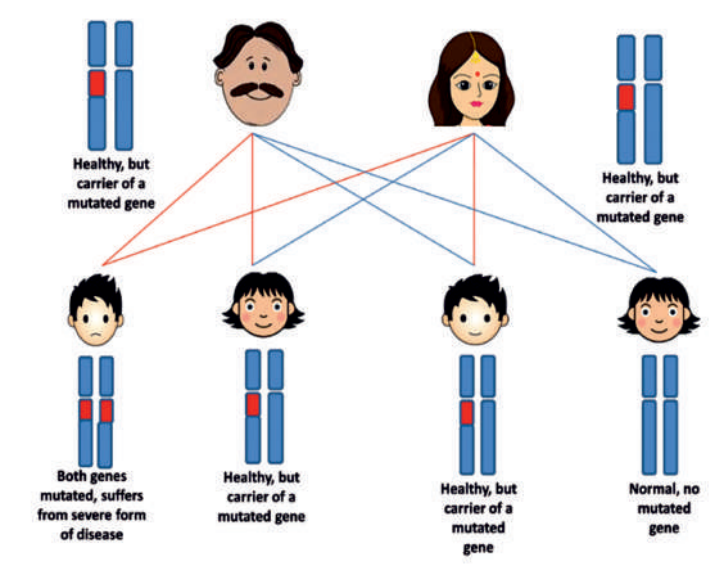
- Usually no
- There is a very small risk for the baby which will be explained to you

Don't be afraid

- It is natural to feel anxious
- This is a test to reduce the chance of having a baby with a life long illness
- If you are feeling worried, speak to us

What Are The Risks With This Result? (both sickle carriers)

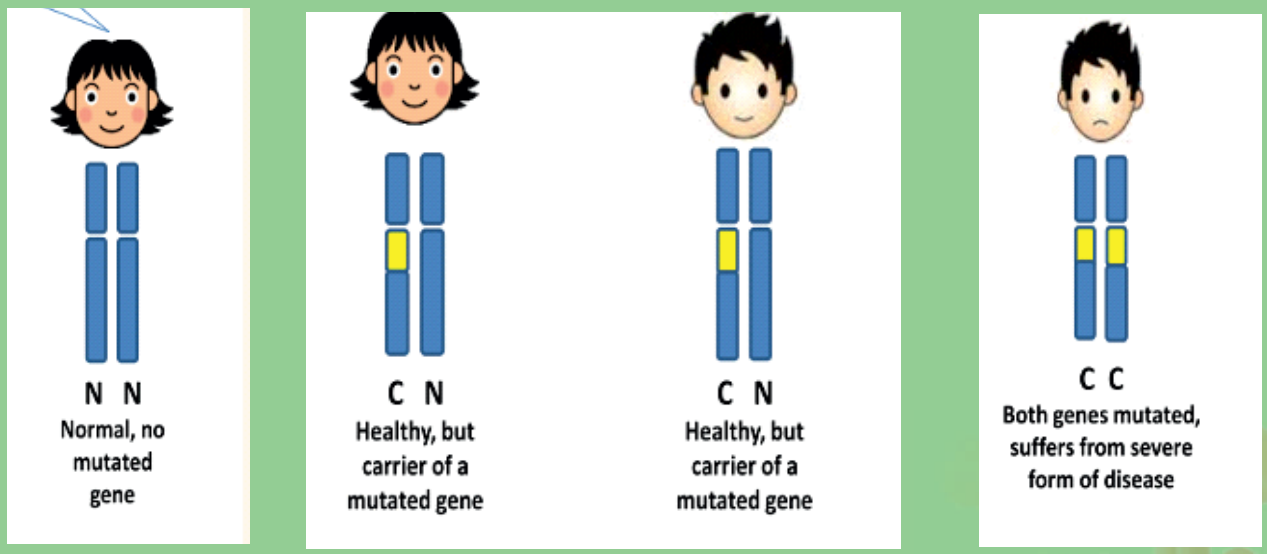
If two sickle cell carriers get married- then they 25% chance of having a child with sickle cell disease.



N= normal gene

C= change or mutated gene

What can be the results of test



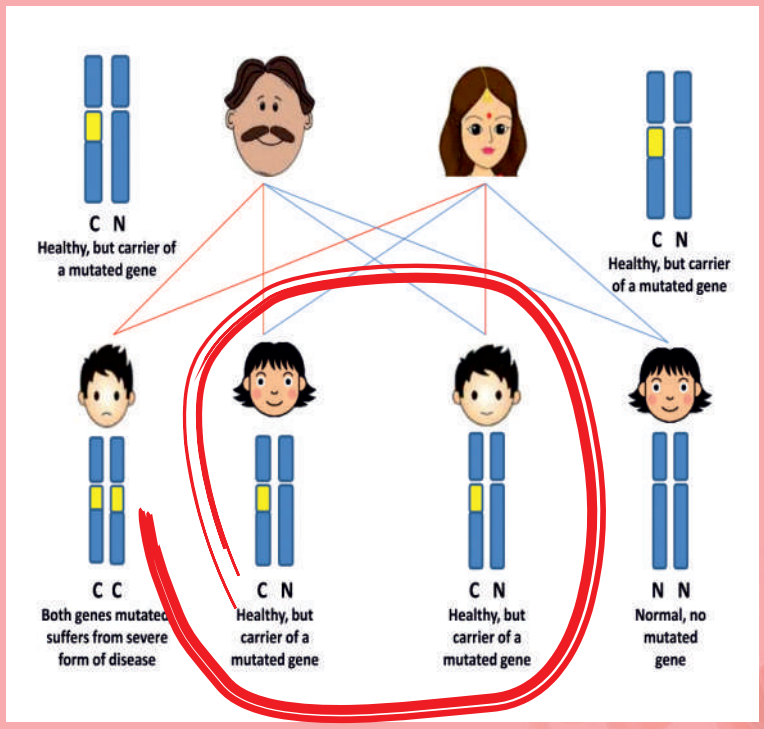
Normal Haemoglobin
HB AA

Sickle Cell trait

Sickle Cell Disease

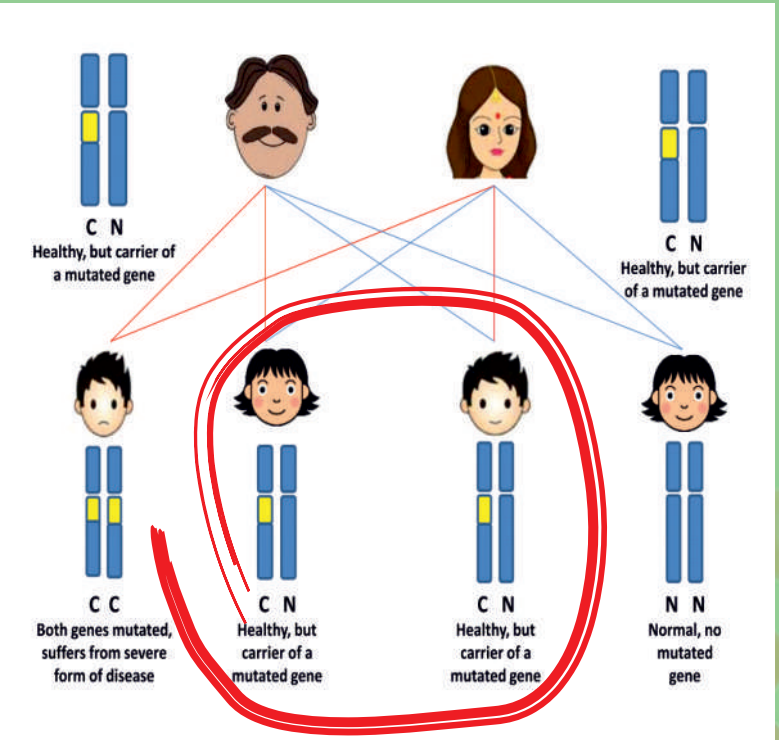
Pre test counseling – Sickle cell carrier state

- If you and your spouse are sickle cell carriers, it is preferable to do prenatal testing to know the baby's sickle status in the first 3 months' of pregnancy
- If the baby has sickle cell trait, pregnancy can be continued as usual

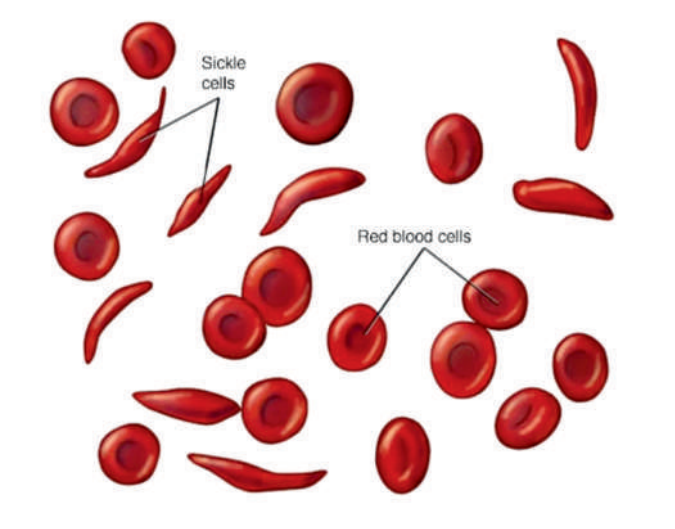


Post test counseling – Sickle cell carrier state

- Your baby has been detected to have a sickle cell carrier state
- Here only one gene is affected and the other is normal
- You can proceed with pregnancy as normal
- After birth, consult your paediatrician for advice regarding long term care and precautions



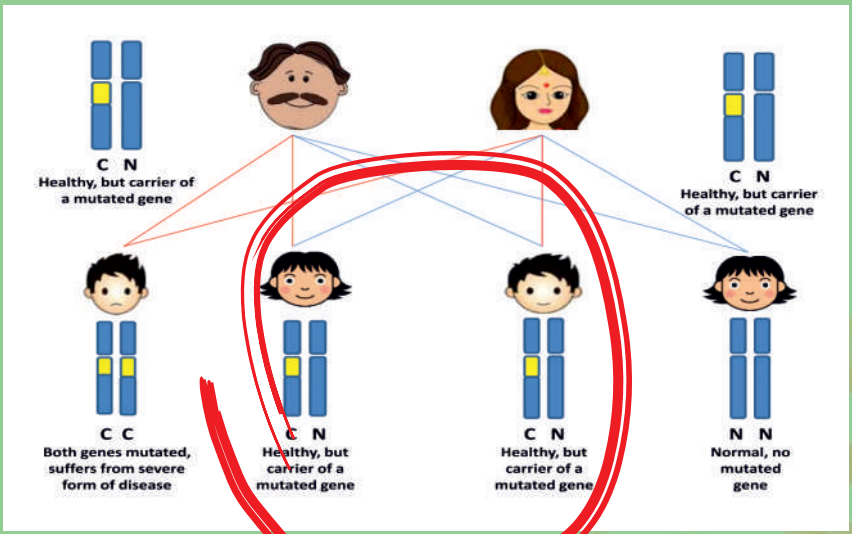
What is sickle cell disease?



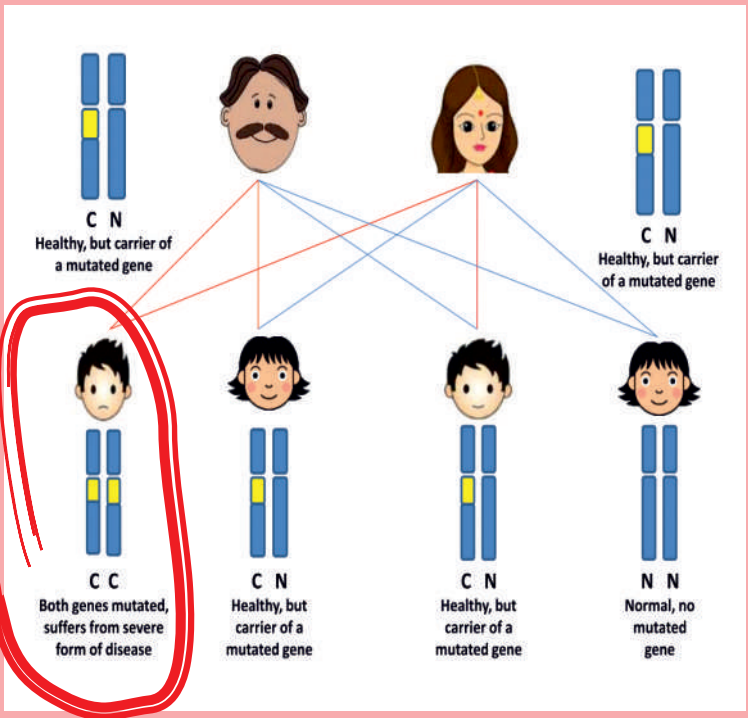
- Sickle cell anaemia is a genetic condition where the haemoglobin molecule inside red blood cells is abnormal.
- The red blood cells carrying sickle-haemoglobin become sticky and deformed (C shaped)
- This can block small blood vessels causing severe pain, and organ damage including stroke.
- These red cells also get destroyed early resulting in anaemia

Post test antenatal counselling when fetus is diagnosed as HbSS

- However, if you decide to proceed with this pregnancy, consult your paediatrician as soon as the child is born for advice regarding long term care and precautions



Post test counseling – Sickle cell disease

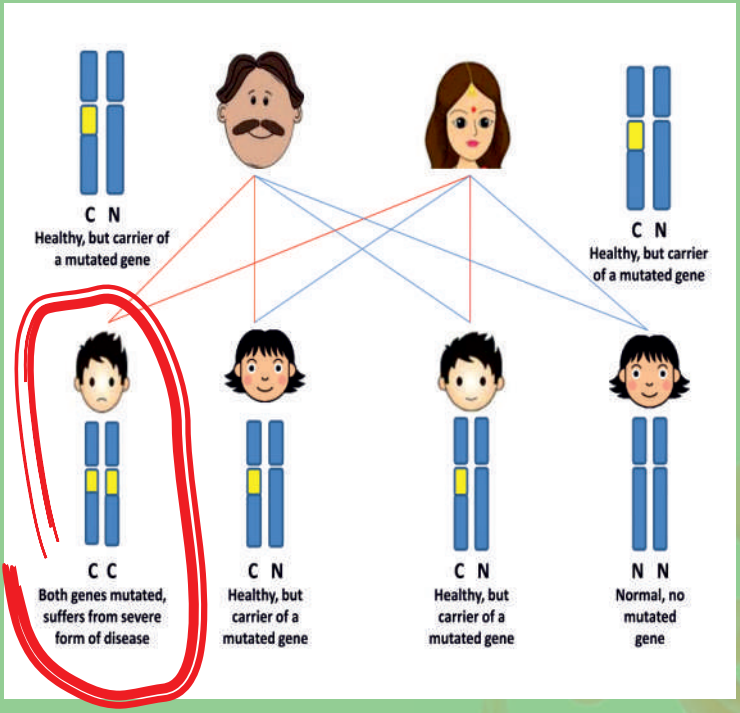


- Your fetus has been detected to have sickle cell disease
- As we discussed earlier, this is a chronic lifelong disease which needs specialised care
- There is an option to do medical termination of pregnancy before 20 weeks of pregnancy

Pre test counseling – Sickle cell carrier state

- If you and your spouse are sickle cell carriers, it is preferable to do prenatal testing to know the baby's sickle status in the first 3 months' of pregnancy

- If the baby has sickle cell disease, then medical termination of pregnancy is allowed, to prevent the birth of the child with a lifelong illness



What does it mean if the baby has Sickle cell disease?

- If you decide to continue with pregnancy,
- This baby will need regular checks to prevent Sickle cell crisis and complications
- The child should be on lifelong medications with Hydroxyurea and penicillin and special vaccinations to prevent infections
- The child may also need blood transfusions and at times exchange transfusions during periods of crisis
- This baby can also have life threatening complications like stroke, severe anemia, risk for clotting etc.

Module Part - IV

Antenatal visits for lady with Sickle Cell Trait

Antenatal care for sickle cell carrier women

- You are a sickle cell carrier
- Your spouse is not a sickle or thalassemia carrier and you have decided to proceed with this pregnancy
- For a sickle carrier, pregnancy and delivery will be similar to any other person without sickle carrier status
- However, you should avoid – extremes of weather, dehydration, strenuous exercise etc.

Module Part - V

Antenatal care for lady with Sickle Cell Disease

Antenatal care for lady with sickle cell disease

- You have sickle cell disease (HbSS)
- You have been on treatment with hydroxy urea and penicillin prior to being pregnant
- You have been counseled regarding the risk of transmitting this gene to your offspring
- Your spouse has been tested and is not a carrier state
- In addition to regular checkup with your gynecologist, you should inform this to your hematologist as well
- **Points to remember**
  1. Complete your vaccination status – hepatitis B, pneumococcal vaccine
  2. Complete blood count
  3. Viral markers – HIV, Hepatitis B, Hepatitis C
  4. Stop Hydroxyurea before becoming pregnant. It may be restarted after the first trimester
  5. Aspirin and Folic acid are required during pregnancy
  6. For pain crisis you can take paracetamol. Other drugs should be taken only after consulting your doctor
- Most women with sickle cell disease do not have any major issues during pregnancy
- Crisis can be triggered during pregnancy
- Avoid dehydration – take more water than usual
- There is a risk of clot in veins during pregnancy if you have sickle cell disease. Keep walking and exercising
- Regular BP, Urine protein measurements are crucial during pregnancy
- Normal delivery is possible. Some patients with sickle cell disease may been caesarean section
- Ask your ASHA to help you during this period
- Pregnant women have a higher risk of developing blood clots in the legs (venous thrombosis) compared with women who are not pregnant.
- There is also a higher chance that you may get pre-eclampsia (a condition of high blood pressure and protein in the urine) in later pregnancy. The blood pressure and urine should be checked regularly.
- Sickle cell disease may also affect your baby's growth because it can affect how your placenta works.

- Check with your doctors about what is best for you and your baby.
- Eat well during pregnancy
- Folic acid tablets are to be taken regularly
- Iron tablet may not be required for sickle cell disease patients who are pregnancy
- Follow all regular ultrasound tests and blood tests during pregnancy
- **Early labour may happen;** you will need to check with your obstetrician about this. You may need to have a caesarean section.
- **<u>Care during crisis</u>**
    - Sickle cell crisis should be prevented as much as possible
    - However, if you develop a crisis, you may need admission
    - In case of high fever, severe pain, difficulty in breathing, giddiness, difficulty in moving arm/leg, difficulty in vision reach emergency
    - You will be started on drips, oxygen, medicines for pain, antibiotics, blood thinners etc as needed

Care during delivery for lady with sickle cell disease

- Insist on a hospital delivery. Avoid home delivery as far as possible
- Normal delivery should be possible in most cases. If there are any other issues for the mother or baby, your doctor may recommend caesarean section
- During delivery – you need to keep yourself hydrated
- You may need drip, oxygen and pain medicines
- Blood transfusion may be required during delivery
- Your baby will be attended to by a doctor during delivery

Care after delivery for lady with sickle cell disease

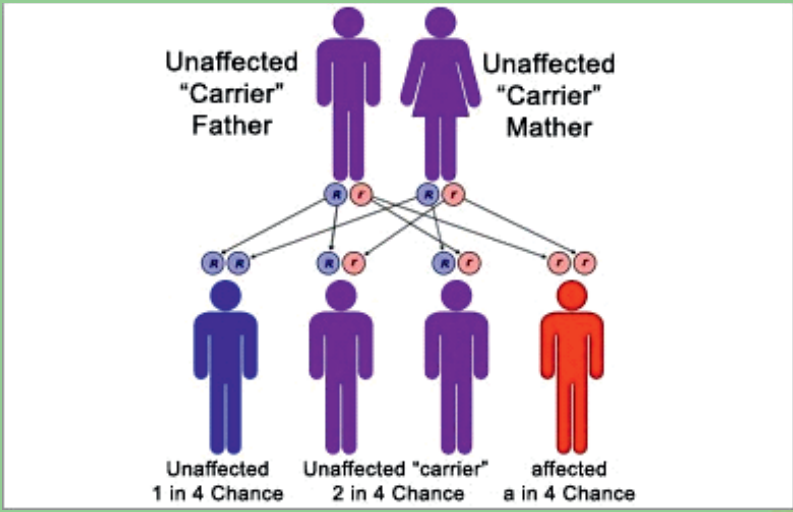
- Be well hydrated
- Start moving as early as possible. This helps reduce the risk of clot formation in the legs
- Start breast feeding your baby
- Continue calcium, folic acid as advised by your doctor
- Contraception can be followed as advised by your doctor
- Contraception after sickle cell disease delivery
- Levonorgestral IUD is contraceptive of choice.
- Other options are progesterone only pill and Depomedroxy progesterone acetate.
- Combination birth control and Copper containing IUD are less preferable.

Module Part - VI

Care of a newborn with Sickle Cell Trait

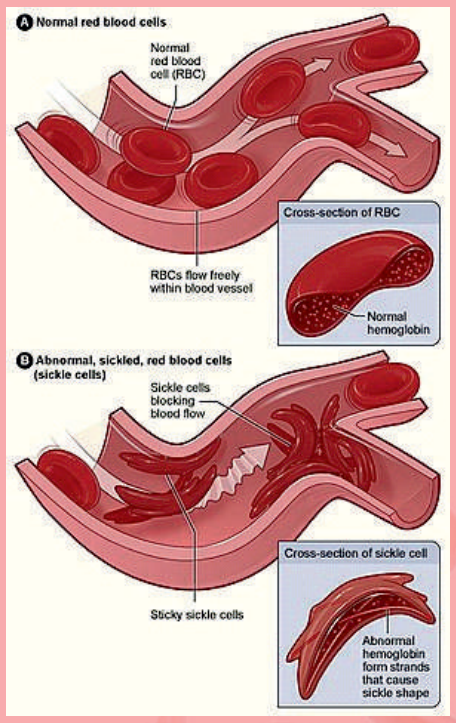
What causes sickle cell disease?

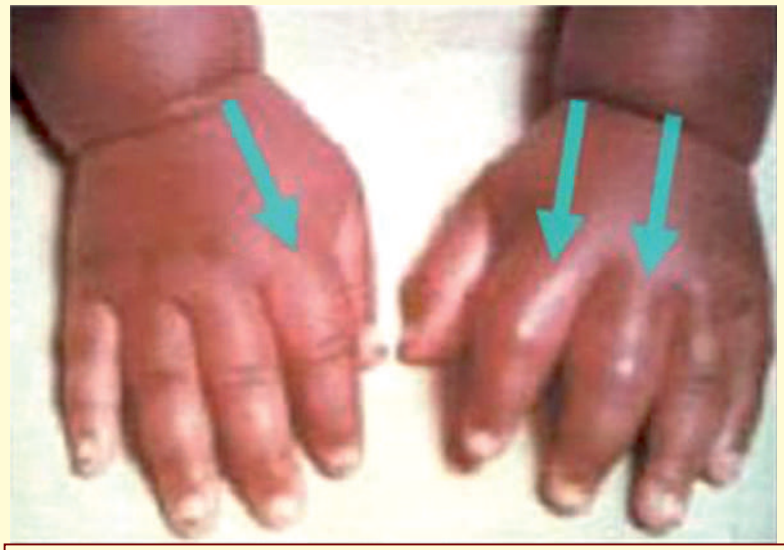
- Sickle cell disease is an inherited condition
- The gene for the globin chain is different (similar to how people have blue-coloured eyes or brown hair)
- It is inherited from parents who have sickle cell trait
- Carriers are usually not symptomatic and can be detected only when screened



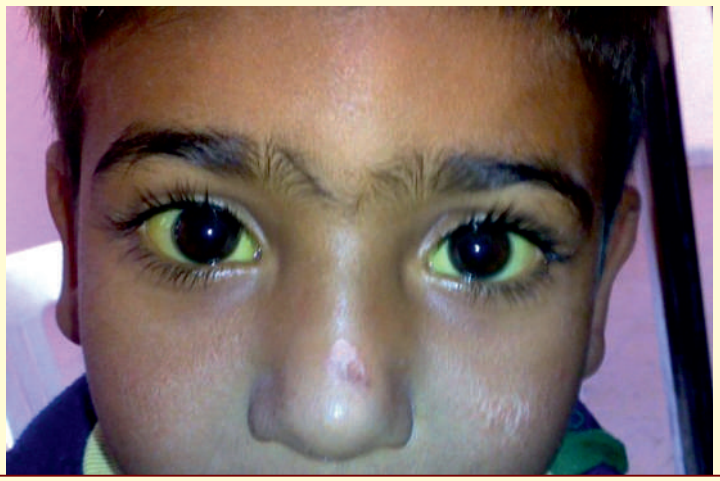
What are the symptoms of sickle cell anemia?

- Sickle cell anemia patients (HbSS) disease are symptomatic from early life
- They present with occlusion of small blood vessels in hands, feet, chest etc
- The early signs are swelling of hands and feet, anemia (paleness of body), repeated fever, fatigue, repeated infections, poor growth, repeated episodes of jaundice etc.





Crisis
Painful swelling of hands and feet



Recurrent episodes of jaundice

Pallor
Child's hands look pale compared to yours

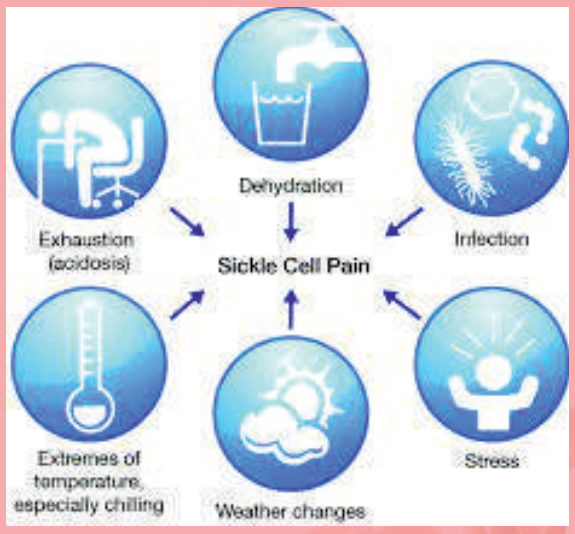
What is a crisis?

- Many children and adults develop what is called a pain crisis.
- During this period, they present to the hospital with pain in the back, legs, chest etc.
- **What triggers pain?**

  Change of weather, diarrhea, fever, dehydration, infection, stress, exhaustion

How can you identify sickle cell anemia crisis?

- **Crisis can present as**
  1. Hand-foot syndrome- swelling and pain of hands and feet
  2. Acute chest syndrome – cough, difficulty in breathing
  3. Eye – can affect blood vessels in eye and reduce vision
  4. Stroke – can affect blood vessels to brain and cause stroke



My child has sickle cell disease. What should I do?

- Sickle cell disease is a chronic condition
- But the good news is that children can be managed well if they are under the care of a doctor
- These children can do well in life, finish their studies, take up a job and get married and have kids as well.
- So, by focusing on good medical care, we can provide them the “healthiest life possible”

How should I take care of my child?

- Your child needs additional support and care compared to any other child
- This includes
  1. Vaccination
  2. Medication
  3. Routine follow-up visits with the doctor
  4. Possibility for curative treatment

What are the vaccinations that need to be given in sickle cell anemia patients?

- All vaccines provided by GOI – vaccination program

In addition

- Meningococcal vaccine
- Pneumococcal vaccine – Conjugate + Polysaccharide
- H. Influenza B vaccine
- Hepatitis B vaccine , Typhoid vaccine is recommended
- COVID vaccine is advised as per age
- Yearly Influenza vaccine advised

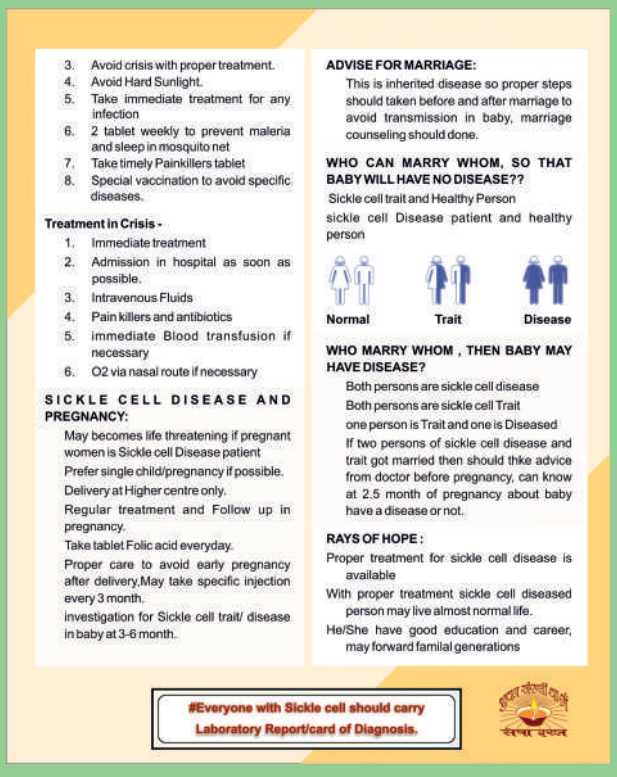
What are the vaccination schedule to be followed? National Immunization schedule

For Infants		
BCG	At birth or as early as possible till one year of age	0.1ml (0.05ml until 1 month age)
Hepatitis B - Birth dose	At birth or as early as possible within 24 hours	0.5 ml
OPV-0	At birth or as early as possible within the first 15 days	2 drops
OPV 1, 2 & 3	At 6 weeks, 10 weeks & 14 weeks (OPV can be given till 5 years of age)	2 drops
Pentavalent 1, 2 & 3	At 6 weeks, 10 weeks & 14 weeks (can be given till one year of age)	0.5 ml
Rotavirus#	At 6 weeks, 10 weeks & 14 weeks (can be given till one year of age)	5 drops
IPV	Two fractional dose at 6 and 14 weeks of age	0.1 ml
Measles /MR 1st Dose$	9 completed months-12 months. (can be given till 5 years of age)	0.5 ml
JE - 1**	9 completed months-12 months.	0.5 ml
Vitamin A (1st dose)	At 9 completed months with measles-Rubella	1 ml (1 lakh IU)

Take your child to a hospital if he/she has

- Repeated fever
- Repeated infections/pneumonia
- Repeated anaemia – needs blood transfusions
- Recurrent episodes of jaundice
- Repeated bouts of pain anywhere in the body
- Swelling of hands, feet or joint pain
- Ulcers in legs
- Is not growing well
- Consult the nearest medical officer or paediatrician

Counselling for SS patients



3. Avoid crisis with proper treatment.
4. Avoid Hard Sunlight.
5. Take immediate treatment for any infection
6. 2 tablet weekly to prevent maleria and sleep in mosquito net
7. Take timely Painkillers tablet
8. Special vaccination to avoid specific diseases.

Treatment in Crisis -

1. Immediate treatment
2. Admission in hospital as soon as possible.
3. Intravenous Fluids
4. Pain killers and antibiotics
5. immediate Blood transfusion if necessary
6. O2 via nasal route if necessary

SICKLE CELL DISEASE AND PREGNANCY:

May becomes life threatening if pregnant women is Sickle cell Disease patient
Prefer single child/pregnancy if possible.
Delivery at Higher centre only.
Regular treatment and Follow up in pregnancy.
Take tablet Folic acid everyday.
Proper care to avoid early pregnancy after delivery.May take specific injection every 3 month.
investigation for Sickle cell trait/ disease in baby at 3-6 month.

ADVISE FOR MARRIAGE:

This is inherited disease so proper steps should taken before and after marriage to avoid transmission in baby, marriage counseling should done.

WHO CAN MARRY WHOM, SO THAT BABY WILL HAVE NO DISEASE??

Sickle cell trait and Healthy Person
sickle cell Disease patient and healthy person

Normal Trait Disease

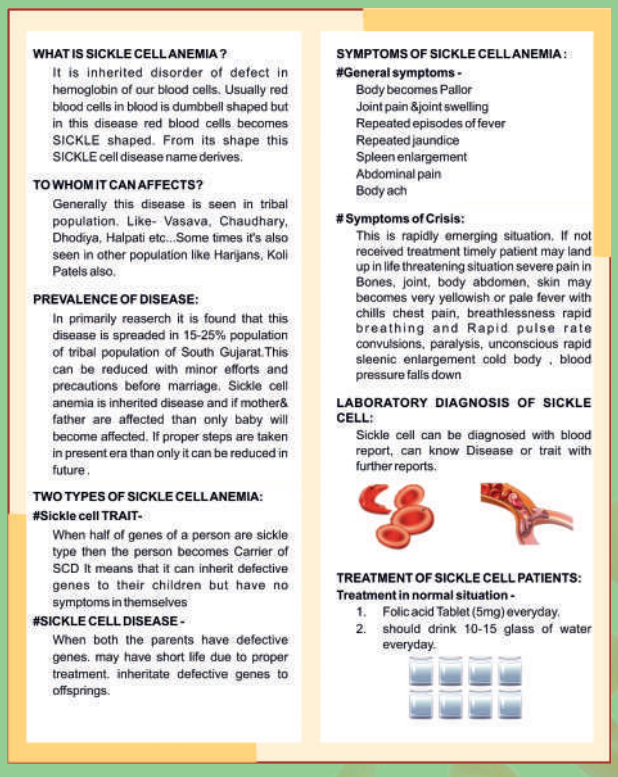
WHO MARRY WHOM , THEN BABY MAY HAVE DISEASE?

Both persons are sickle cell disease
Both persons are sickle cell Trait
one person is Trait and one is Diseased
If two persons of sickle cell disease and trait got married then should thke advice from doctor before pregnancy, can know at 2.5 month of pregnancy about baby have a disease or not.

RAYS OF HOPE :

Proper treatment for sickle cell disease is available
With proper treatment sickle cell diseased person may live almost normal life.
He/She have good education and career, may forward familal generations

#Everyone with Sickle cell should carry Laboratory Report/card of Diagnosis.



WHAT IS SICKLE CELL ANEMIA ?

It is inherited disorder of defect in hemoglobin of our blood cells. Usually red blood cells in blood is dumbbell shaped but in this disease red blood cells becomes SICKLE shaped. From its shape this SICKLE cell disease name derives.

TO WHOM IT CAN AFFECTS?

Generally this disease is seen in tribal population. Like- Vasava, Chaudhary, Dhodiya, Halpati etc...Some times it's also seen in other population like Harijans, Koli Patels also.

PREVALENCE OF DISEASE:

In primarily reaserch it is found that this disease is spreaded in 15-25% population of tribal population of South Gujarat.This can be reduced with minor efforts and precautions before marriage. Sickle cell anemia is inherited disease and if mother& father are affected than only baby will become affected. If proper steps are taken in present era than only it can be reduced in future.

TWO TYPES OF SICKLE CELL ANEMIA:

#Sickle cell TRAIT-

When half of genes of a person are sickle type then the person becomes Carrier of SCD It means that it can inherit defective genes to their children but have no symptoms in themselves

#SICKLE CELL DISEASE -

When both the parents have defective genes. may have short life due to proper treatment. inheritate defective genes to offsprings.

SYMPTOMS OF SICKLE CELL ANEMIA :

#General symptoms -

Body becomes Pallor
Joint pain &joint swelling
Repeated episodes of fever
Repeated jaundice
Spleen enlargement
Abdominal pain
Body ach

 # Symptoms of Crisis:

This is rapidly emerging situation. If not received treatment timely patient may land up in life threatening situation severe pain in Bones, joint, body abdomen, skin may becomes very yellowish or pale fever with chills chest pain, breathlessness rapid breathing and Rapid pulse rate convulsions, paralysis, unconscious rapid sleenic enlargement cold body , blood pressure falls down

LABORATORY DIAGNOSIS OF SICKLE CELL:

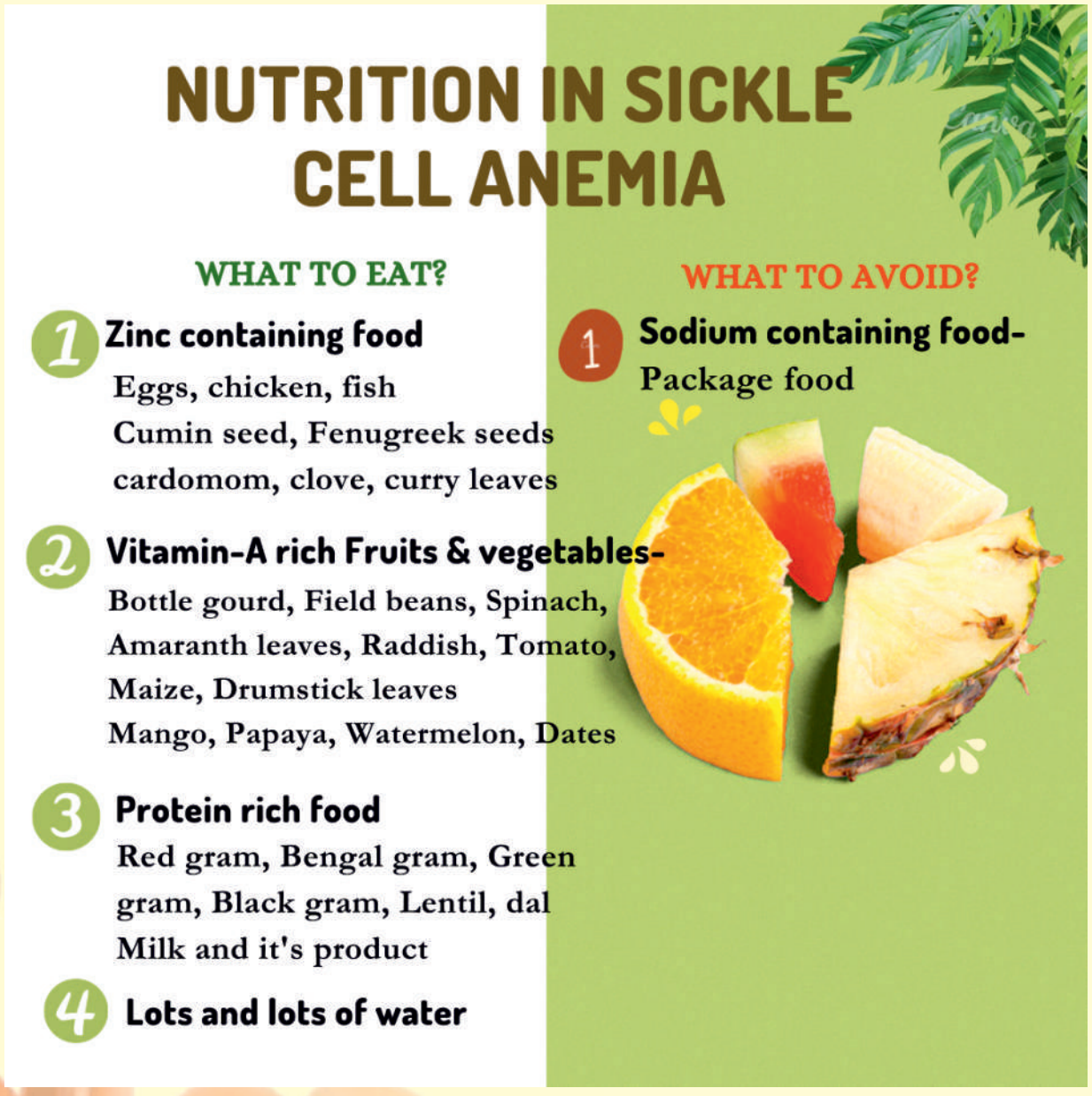
Sickle cell can be diagnosed with blood report, can know Disease or trait with further reports.

TREATMENT OF SICKLE CELL PATIENTS:

Treatment in normal situation -

1. Folic acid Tablet (5mg) everyday.
2. should drink 10-15 glass of water everyday.

Nutrition in Sickle cell anemia



सत्यमव जयते

Government of India

Ministry of Tribal Affairs & Ministry of Health & Family Welfare