सत्यमेव जयते

Ministry of Health & Family Welfare
Government of India

National Sickle Cell Anaemia Elimination Mission 2023

Training Module for Staff Nurses

June 2023

CONTENT

CHAPTER 1
OVERVIEW OF SICKLE CELL DISEASE IN INDIA

Sickle Cell Disease (SCD) is a genetic disorder where the red blood cells have an abnormal half-moon shape. It not only causes anaemia but also pain, reduced growth, and affects many organs like lungs, heart, kidneys, eyes, bones, and brain.

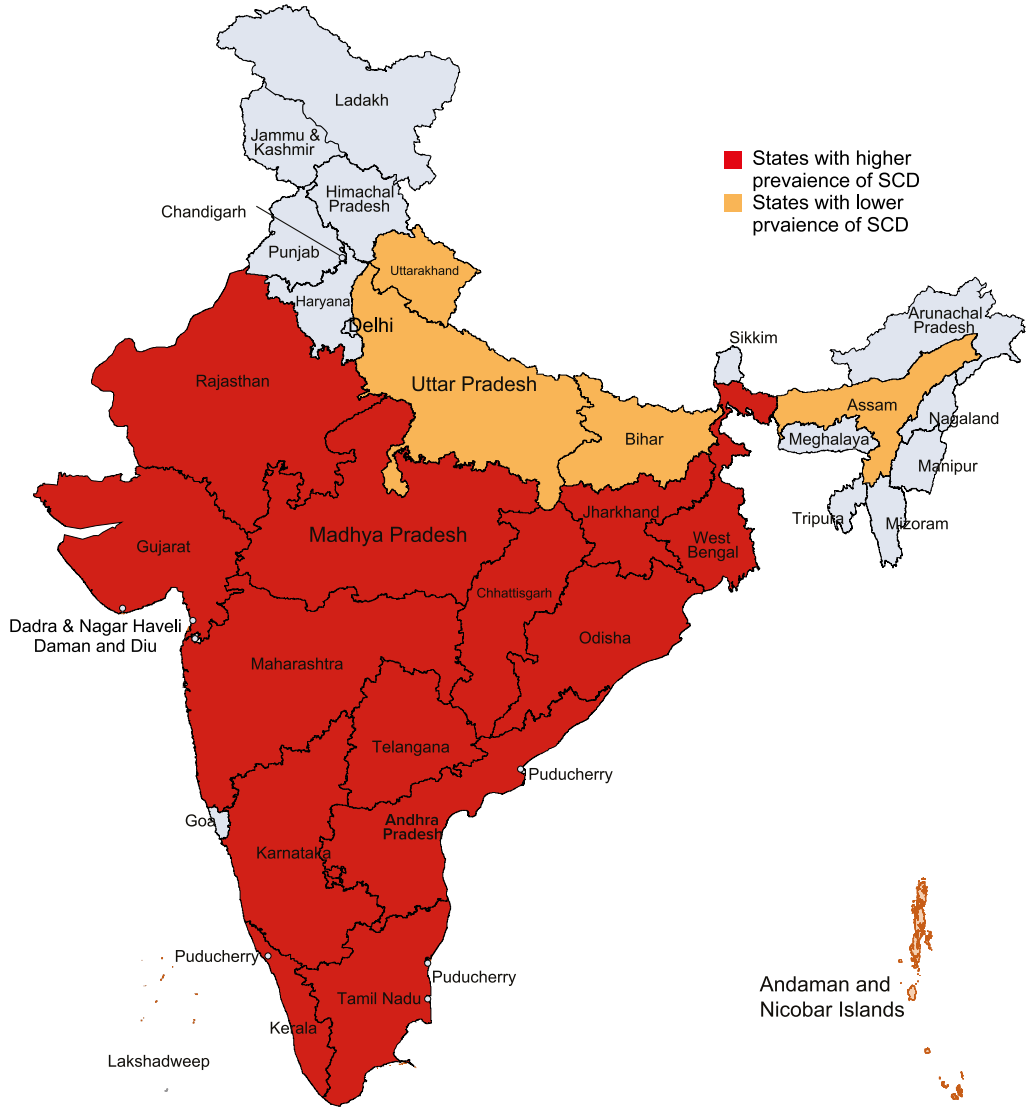
Figure 1: Normal red blood cells can live up to 120 days

Figure 2: Sickled red blood cells live up to 10-20 days

1.1 Burden

States with higher prevalence of Sickle Cell Disease include Gujarat, Rajasthan, Uttarakhand, Maharashtra, Bihar, Jharkhand, Madhya Pradesh, Chhattisgarh, Odisha, West Bengal, Tamil Nadu, Telangana, Andhra Pradesh, Karnataka, Kerala, Uttar Pradesh & Assam. SCD is widespread in many tribal groups of India. About 1 in 86 births among Tribal population have SCD, the prevalence being higher in Central, Western, and Southern India. However now SCD is found across all ethnicities and communities.

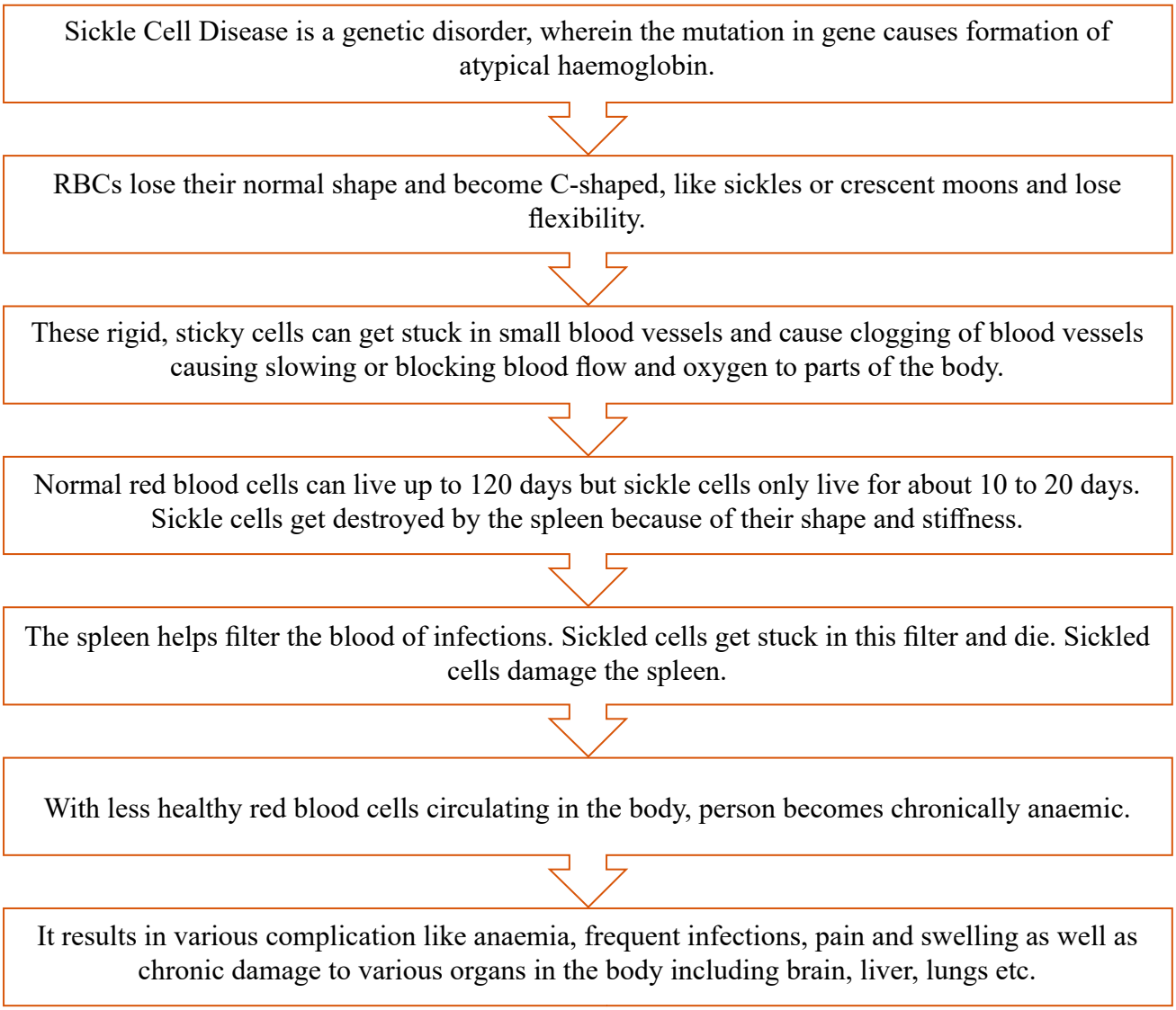
Figure 3: States with prevalence of SCD



1.2 Aetio-pathology of SCD

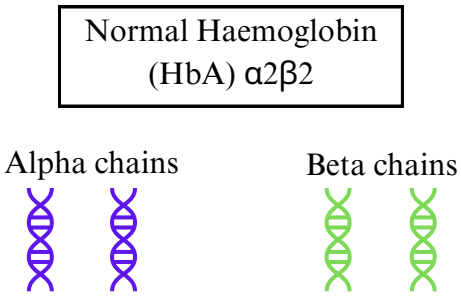
Hemoglobin (Hb) is a protein-based molecule found in the red blood cells (RBC) that carries oxygen in our body and gives blood its red colour. Normal red blood cells are biconcave, have no nucleus and being flexible can easily change shape, which helps them to fit and move easily through smallest blood vessels called capillaries. The following flowchart shows the aetiopathogenesis of Sickle cell disease.

Figure 4: Aetiology of SCD

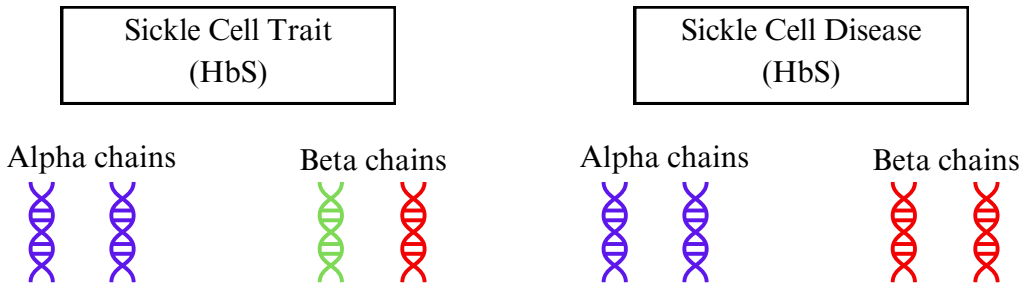


1.3 Types of Sickle cell

As a Staff Nurse it is important for you to know the types and transmission of Sickle Cell Disease and Trait from one generation to another. Normal human hemoglobin (Hemoglobin A or HbA), also known as adult hemoglobin, (Hemoglobin A or $\alpha 2\beta 2$) consists of two subunits of beta globin and two subunits of alpha globin. These two genes have to function normally and in tandem to produce normal hemoglobin in human children and adults.

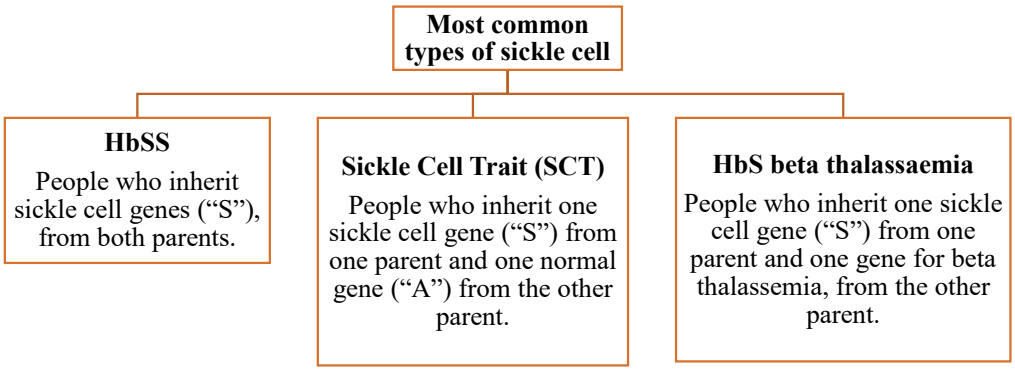


When faulty haemoglobin replaces normal haemoglobin (HbA), the person can be a sickle cell carrier or have sickle cell disease. Sickle haemoglobin (HbS) is result of a point mutation in the beta globin chain. If only one subunit of beta globin is affected, the person has trait, and if both are affected, the person has sickle cell disease.



Patients with sickle cell trait inherit HbS from one parent and HbA from the other, making them heterozygous. Patients with sickle cell disease inherit two genes that code for HbS from both parents, making them homozygous.

Figure 5: Most common types of Sickle Cell

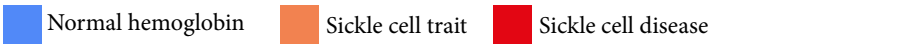


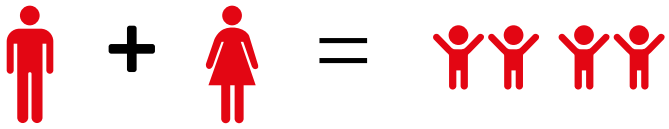
People with SCT usually do not have any of the signs of the disease and live a normal life, but they can pass the trait on to their children. Additionally, there are a few, uncommon health problems that may potentially be related to sickle cell trait like exertion and exercise intolerance, etc.

1.4 Inheritance of Sickle cell

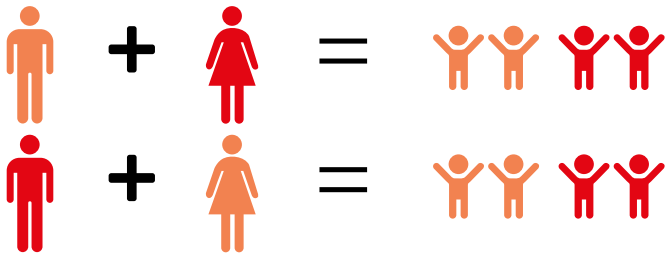
As a Staff Nurse it is your responsibility to counsel a diseased and carrier individual appropriately to control the transmission of the genes to the upcoming generation. A pictorial depiction of different combinations of parents' disease status and the probability of the children getting affected is as below:

Figure 6: Inheritance patterns in different carrier states

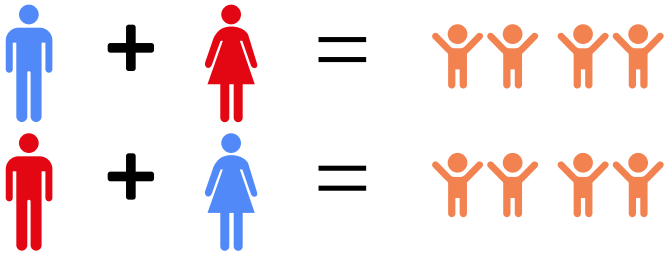




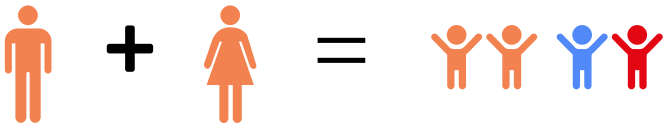
If both parents have sickle cell disease, there is a 100% chance that their children will be born with the disease



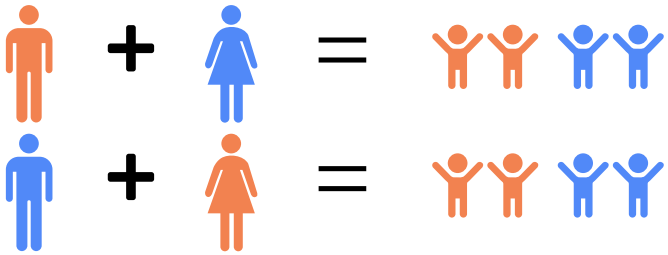
If one parent has sickle cell trait and the other has sickle cell disease, then children have a 50% chance of being diseased and 50% of being carriers



If one parent is normal and the other has sickle cell disease, then children have a 100% chance of being carriers



If both parents have sickle cell trait, their children have a 25% chance of being diseased, a 25% chance of being normal, and a 50% chance of being carriers



If one parent has sickle cell trait and the other is normal, then children have a 50% chance of being normal and 50% chance of being carriers

1.5 Signs and symptoms

Patients suffering from Sickle Cell Disease experience a wide range of symptoms. While some start showing signs and symptoms early in life, some develop the symptoms later in life. Being a Staff Nurse, you should be aware of the presenting symptoms to help identify the cases and manage them at facility level. Following are the common symptoms experienced by an individual suffering from SCD.

- **Patient may be anaemic as s**ickle cells break apart easily and die, leaving a shortage of red blood cells.
- Patient may feel **fatigued** due to lack of oxygen
- Patients may experience **periodic episodes of extreme pain**, called pain crises. Pain develops when sickle-shaped red blood cells block blood flow through tiny blood vessels of chest, abdomen and joints. The pain varies in intensity and can last for a few hours to a few days. Cases with severe pain needs to be referred to hospital.
- Owing to vaso-occlusion, some patients may also have **chronic pain**, resulting from bone and joint damage, ulcers, and other causes.
- Sickle-shaped RBCs may block blood circulation and cause s**welling of hands and feet**
- Sickle cells causing vaso-occlusion can damage the spleen, increasing **vulnerability to infections**.
- Shortage of healthy red blood cells can **slow growth** in infants and children and **delay puberty** in teenagers.
- Tiny blood vessels in the eyes may become plugged with sickle cells damaging retina and leading to **vision problems**.
- Patients might present with **bouts of breathlessness.** Breathlessness may be precipitated on exertion and heavy work
- Persons suffering from sickle cell anaemia have increased risk of having a **stroke**

1.6 Crisis/ Complications in Sickle Cell Disease

Some patients experience complications and may require hospitalization depending upon the severity of the symptoms. In such cases you should be able to identify the crisis early, provide the patient with basic medical care and refer the patient to the higher facility for better treatment.

- A **vaso-occlusive crisis** occurs when the microcirculation is obstructed by sickled red blood cells resulting in ischemic injury. The patient will be experiencing, any of the following:
    - Pain, usually affecting bones such as femur, tibia and lower vertebrae.
    - Dactylitis i.e., painful and swollen hands and/ or feet
    - Painful abdomen.
    - The spleen may undergo auto-infarction and is often not palpable beyond 6 years of age.
    - Inability to concentrate urine.
    - Acute chest syndrome
    - Retinal haemorrhages
    - Priapism,
    - Pain near hip join and difficulty in walking

- Cerebrovascular accidents.
- Leg ulcer

- **Acute chest syndrome:** This requires urgent admission; oxygen support, antibiotics, intravenous fluids, bronchodilators and use of steroids may be of benefit. This is a type of vaso-occlusive crisis affects the lung and presents with:
  - Chest pain
  - Cough
  - Tachypnoea
  - Dyspnoea
  - Hypoxemia
  - Fever or a new pulmonary infiltrate.
- **Sequestration crisis:** This is due to sickled cells that block splenic outflow, leading to the pooling of peripheral blood in the engorged spleen resulting in splenic sequestration.
- **Aplastic crisis:** Aplastic crisis can occur when the bone marrow stops producing red blood cells. This is most commonly seen in patients with infection or folate deficiency. This is usually self-limited and may follow viral infections of which parvovirus B19 is the most commonly implicated. Usually only supportive care and occasionally packed red blood cell transfusions are required.

1.7 Social and economic implications of Sickle Cell Disease (SCD)

Suffering from SCD can contribute to

- Clinical symptoms
- Depressive symptoms
- Absenteeism
- Deterioration in productivity.

Suffering from SCD had indirect effect on the socio-economic condition of the families and wider society.

- The first is due to lost productivity because of parents' missing days of work to look after their children.
- The second is the future loss to the economy as sickle cell disease deaths mainly occur in children and young adults causing early deaths that erodes the future workforce.
- Given the association with health care costs due to long term management, and aforementioned associated factors, this may also lead to financial hardship for population subgroups of middle and lower economic status.

CHAPTER 2
NATIONAL SICKLE CELL ANAEMIA ELIMINATION MISSION

SCD has a significant macroeconomic effect on an individual, their family as well as on the nation. Financial implications include both on the nation and the individual. Direct costs are those that are met by the healthcare system; for sickle cell disease, these include:

- Cost of screening
- Primary and emergency care visits
- Cost of drugs
- Hospitalizations
- Blood transfusions
- Bone marrow transplants
- Other out-of-pocket expenditure borne by the patient.

To overcome the physical, psychological and economic impact of dealing with SCD at individual and national level, the Government of India, launched the Sickle Cell Mission, through the budget announcement in 2023.

2.1 Goal

Eliminate sickle cell disease as a public health problem in India before 2047

2.2 Objectives

1. Provide affordable, accessible care and ensure quality to all SCD patients
2. To reduce the prevalence of SCD

These objectives would be attained through strategies spanning awareness generation, strengthening of screening and testing facilities, strengthening of laboratory services for diagnosis, facilitation of management & treatment, establishing linkages across level of care, inter sectoral convergence towards holistic approach and linkages with social security schemes/ benefit packages.

2.3 Strategic Pillars

The three strategic pillars for SCD elimination are:

I. Primary prevention strategies:

- Primary prevention strategies focus on awareness generation and pre-marital and pre-conceptional counselling to prevent the conception of a child with homozygous genotype.
- Prevention requires setting up genetic counselling and testing interventions in high prevalence districts to prevent sickle cell disease in the offspring. Genetic counselling and health promotion activities can lead to substantial reduction in the number of children born with the disease.
- Widespread community involvement and support are essential as there are existing diversity of cultures and opinions about a number of issues relevant to genetics, such as human reproduction issues.

II. Secondary Prevention and Screening:

Secondary prevention focuses on the following components related to early diagnosis and care of sickle cell disease.

- Screening for detection of Sickle Cell Trait to reduce the birth of children affected with Sickle Cell Disease and screening for early detection of sickle cell disease to achieve a reduction in mortality and morbidity with improvement in quality of life of the affected.

III. Holistic management and continuum of care

- Management of persons with sickle cell disease at primary, secondary, and tertiary health care levels
- Advanced diagnostic and treatment modalities at tertiary health care facilities
- Integration with AYUSH
- Patient support system
- Community Adoption
- Rehabilitation

2.4 Scope for Sickle Cell Disease Elimination Program

The Sickle cell elimination program is a part of National Health Mission and focuses on universal population-based screening, prevention, and management of sickle cell disease high prevalent States of India. While in its initial stage, the mission would prioritize its intervention in high prevalence states, the plan would subsequently expand to include all states in phases incrementally.

The program is carried out in a mission mode covering the entire population from zero to forty years. The mission aims to cover 7 crore people with screening, counselling for prevention and care for people with SCD in three years.

The programme would be in integration with existing mechanism and strategies under NHM to ensure utilization of existing resources and also minimizing the duplication of efforts, for example, established platform of Rashtriya Bal Swasthya Karyakram (RBSK), Pradhan Mantri Surakshit Matritva Abhiyan (PMSMA) and Anemia Mukt Bharat to be leveraged to achieve the targets for the Sickle Cell mission. The mission will be further linked to the national program for Prevention and Control of Haemoglobinopathies in India.

2.5 Role of Primary Health Care team in SCD prevention

Primary health care team including Medical Officers, Staff Nurses, Community Health Officers, Auxillary Nurse Midwife, Multi-purpose workers (M/F), Accredited Social Health Activists in Ayushman Bharat-Health and Wellness Centre is associated with all aspects of SCD prevention, control, counselling, and management such as:

- Organizing community awareness events on SCD on a regular basis
- Screening people in remote tribal hamlets through mobile medical units or through dedicated teams
- Conducting facility-based screening
- Conducting opportunistic screening for people attending outpatient services at the Health and Wellness Centre.
- Providing counselling services
- Referral to nearest secondary care facility (CHC/DH)

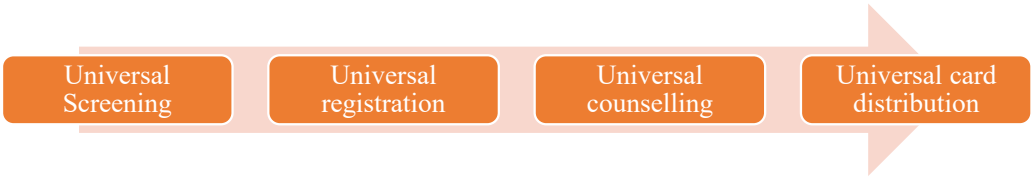
CHAPTER 3
SCREENING AND COUNSELLING FOR PREVENTION

3.1 Screening tests available at AB-HWC

For screening of SCD and SCT the following tests are available at the AB-HWC level. However, the availability of the tests depends on the state. The state may adapt to either of the approaches for determining the status of SCD. You will screen individuals and based on the outcome you will provide support.

- **Option 1:** One step approach - Point of Care test, approved by Government of India will be used as a one-step confirmatory test. This test can be performed by staff with minimal training. The test rapidly distinguishes normal, carrier and sickle cell disease status.
- **Option 2:** Two-step approach- Mass screening / Initial screening using Solubility test. If found positive for Solubility test in field setting then confirmation using Point of Care confirmatory test or HPLC / electrophoresis at higher centers.

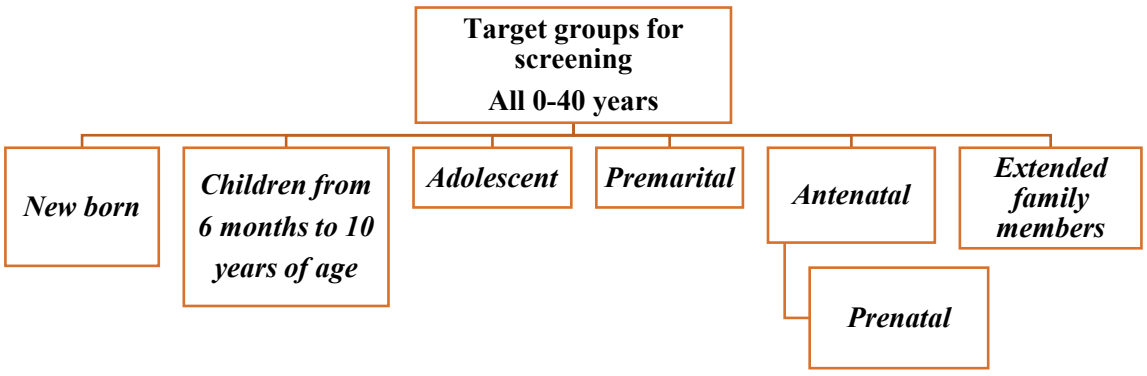
Figure 7: Universal approach



3.2 Screening approaches

To attain a sickle cell free nation, the major intervention is screening and early diagnosis of cases and traits. In order to cover every possible person, you will divide the population residing within your catchment area into 6 groups, having a separate approach for every group.

Figure 8: Target groups for screening



You along with your AB-HWC team will provide screening services at facility level through target group approach and opportunistic screening. You will also conduct monthly outreach sickle cell camps within community to conduct screening and raise awareness. The following table shows the various screening approaches against the target groups:

Table 1: Target groups for screening

Target groups	Setting	Role as a SN
New born	New-born screening shall be conducted at all public health institutions conducting institutional deliveries, in sickle cell endemic areas.	Counsel pregnant mothers belonging to SCD endemic areas to undertake the screening after delivery Collection of Dried Blood Stain (Annexure 2)
Children from 6 months to 10 years of age	Screening of children by RBSK teams either through camps or in Anganwadis / Schools / Ashramshalas / Eklavya Model Residential School (EMRS)	Support the RBSK team in screening children in Anganwadis / Schools / Ashramshalas / Eklavya Model Residential School (EMRS)
Adolescent	At primary care facilities or outreach camps. Rastriya Kishore Swastya Karyakram (RKSK) shall be leveraged for awareness and screening of adolescents	Support RKSK teams in screening adolescents at Adolescent Friendly Health Clinics (AFHCs)
Premarital	At primary care facilities or outreach camps	Screen and counsel all willing couples at facility and during outreach camps
Prenatal	Antenatal screening of all pregnant mothers in sickle cell anaemia affected geographies shall include compulsory screening for SCD along with other tests to detect high risk pregnancy. The Pradhan Mantri Surakshit Matritva Abhiyan (PMSMA) program would be leveraged for this. Prenatal diagnosis to be undertaken at the tertiary care facilities	Screen all pregnant women during Antenatal check-ups at facility and during outreach camps. If found a carrier, her husband too would be tested The couple should be counselled on the pregnancy outcomes and its subsequent management. Refer pregnant woman (8-12 weeks) with SCD to higher facilities to undertake this screening test. Partner screening, at facility, of the antenatal woman who is positive for SCD and SCT
Extended family members	Outreach screening and facility-based camps may be adopted	Counsel and screening family members at facility and during outreach camps facility and during outreach camps

3.3 Sickle cell cards

Every individual who is screened for SCD will be provided a Sickle cell card. The card will show the status of the individual viz, Normal, Carrier or Diseased. The cards are colour coded – blue cards for male and pink cards for female. Based on the card's status the individual will be counselled. The Sickle cell cards will be extensively used for the purpose of counselling by matching the cards of prospective matches. The cards are so designed that can help in counselling the outcome of conception.

The rear side of the card has details on possible outcomes of conception if any person with either sickle cell disease or carrier marries. While matching the cards they should be placed together and held

against the light, the holes coinciding will give the possibility of having Sickle Cell Disease or Trait in the child. There can be five possible outcomes of the pregnancy as discussed in Figure 6. Based on the matching outcomes the following cases should be counselled and referred for Pre-Natal diagnosis:

Table 2: Pregnancies needing Pre-Natal diagnosis

Pre Natal Diagnosis Not Required	***Pre Natal Diagnosis Required.***
If two individuals having sickle cell disease marry, there is a 100% chance that their children will be born with SCD	If a sickle cell disease individual and a sickle cell trait individual marry, there is a 50% chance that their children will be born with the disease and 50% chance that their children will be carriers
If a sickle cell disease individual and a normal individual marry, there is a 100% chance that their children will be born with sickle cell trait.	If two individuals having sickle cell trait marry, their children have 25% chance of being diseased, 25% of being normal and 50% chance of being carriers.
If a sickle cell disease trait and a normal individual marry, their children have 50% chance of being normal and 50% chance of being carriers.	

Figure 9: Sickle cell cards for males

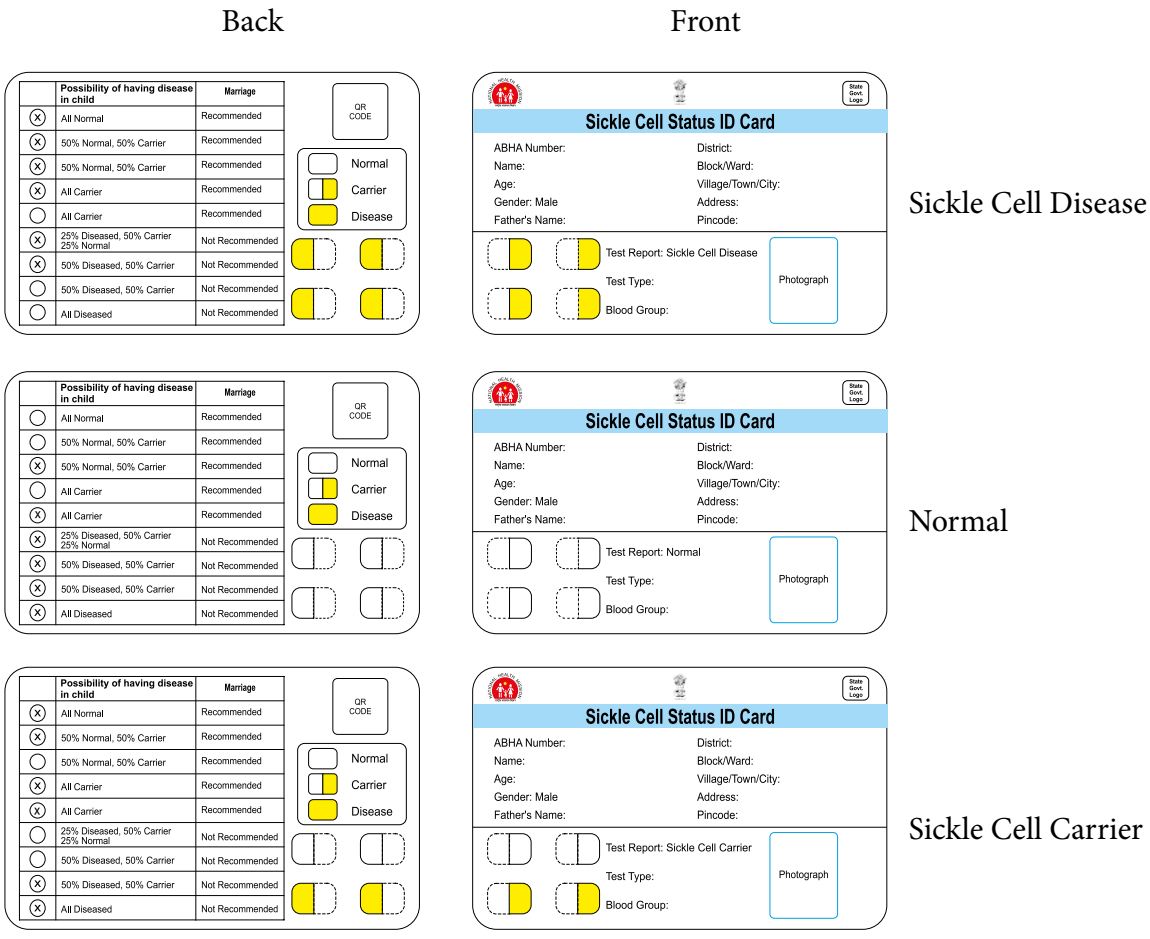


Figure 10: Sickle cell cards for females

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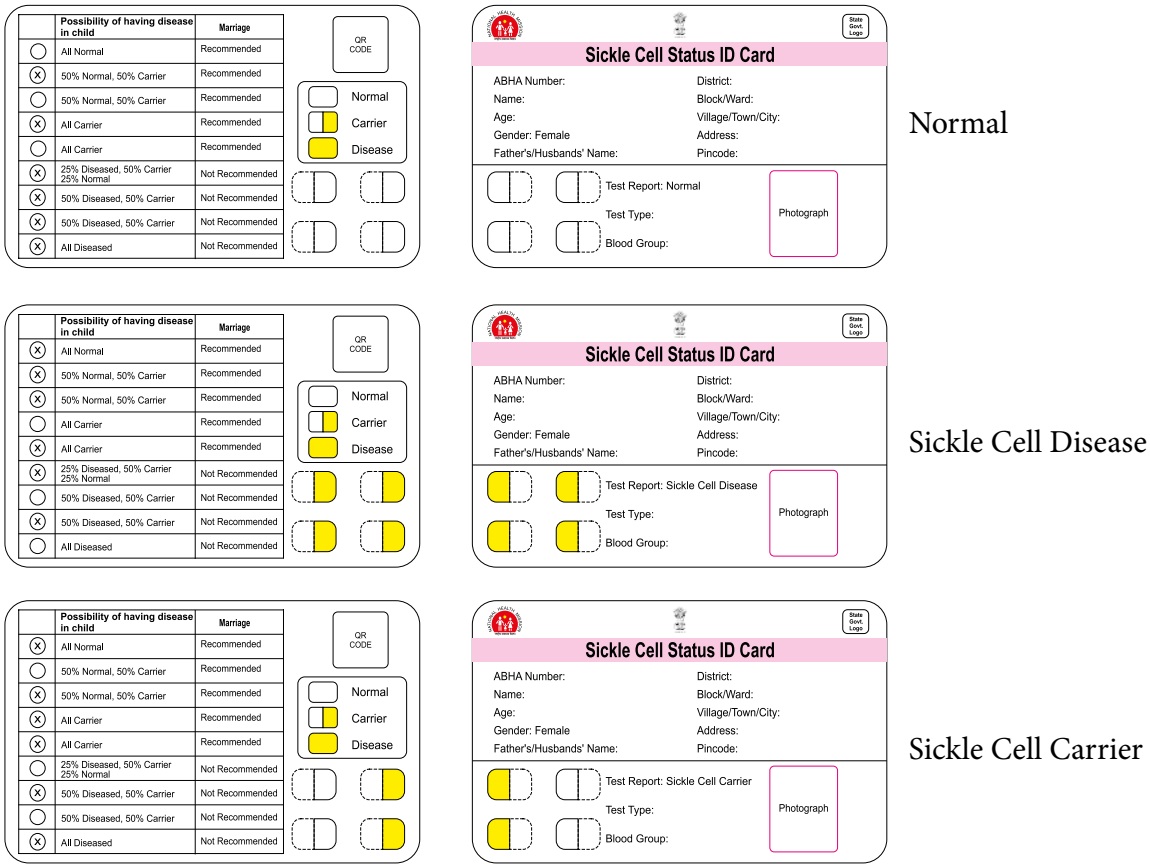


Normal

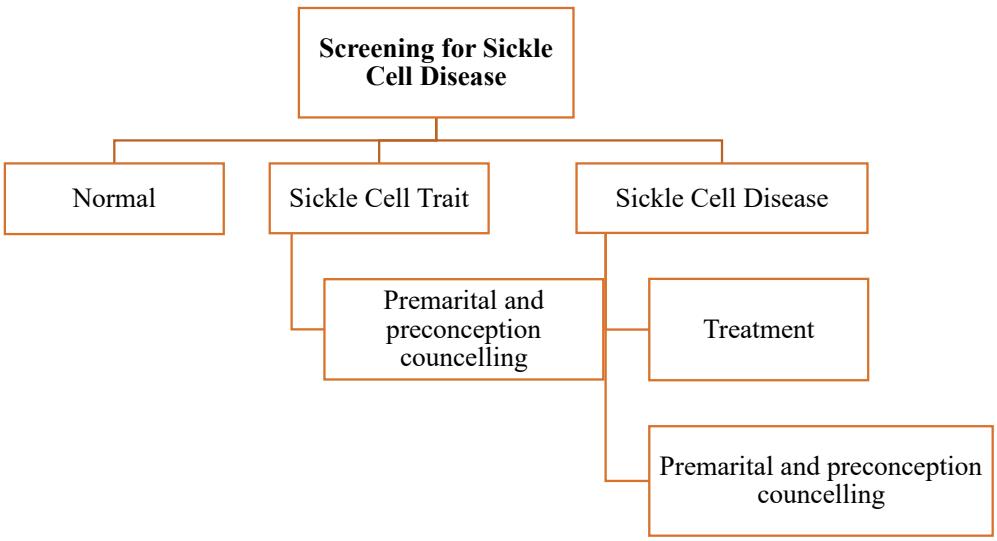
Sickle Cell Disease

Sickle Cell Carrier

3.4 Support to SCD and SCT cases

On identification of cases or traits you will provide required support at the earliest. The following diagram shows the treatment and counselling approach.

Figure 12: Screening outcomes



- ***Registration -*** *Every individual you screen shall be*
  - Registered with an **ABHA ID**
  - Provided with a **Sickle cell card**
  - Detail entered on the **Sickle cell portal** and mobile application
- ***Counselling*** – You will counsel individuals / couples / family to make them aware about sickle cell and the services available and ensure them of the support available at the AB-HWC.
- ***Genetic counselling*** – Once screened you should handle the individuals sensitively and provide counselling. You will use Sickle cell cards extensively for this purpose.
- ***Treatment initiation*** – based on the appearance and severity of the symptoms you will refer the cases to the PHC-MO/UPHC-MO for treatment initiation.
- ***Outreach camps*** **–** Through outreach camps you will extend the services regarding screening, treatment and management of sickle cell disease and trait to the people residing in remote tribal hamlets and other pockets.
- You will also ensure any state specific mass screening camp method which may include outsourcing to appropriate agencies during the outreach camps.

3.5 Prevention and counselling

Your role as a Staff nurse in prevention of SCD at various levels is as follows

Figure 13: Role of SN in prevention and counselling

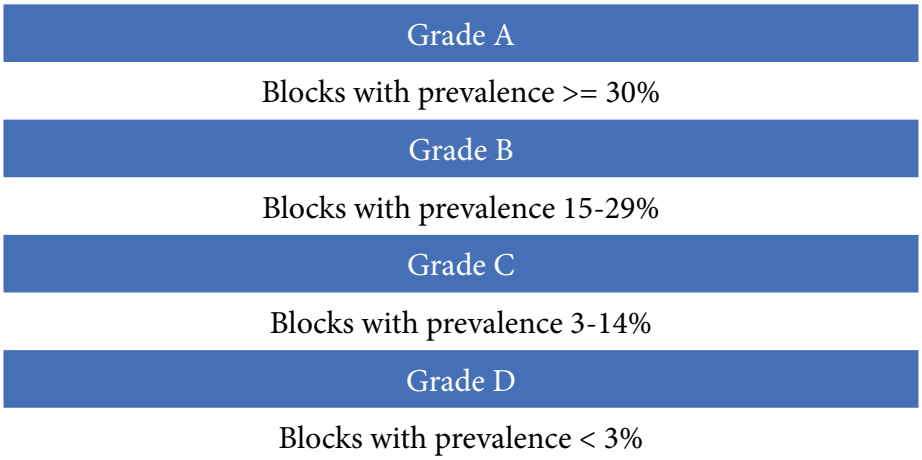
At Community Level	Along with PHC-MO/UPHC-MO, organize regular Sickle cell camps in places with community gathering such as haat bazaar, or fixed day markets Inform ASHAs about the dates and location of Sickle cell camps. ASHAs would mobilize community members to participate Leverage JAS, VHSNC, MAS, CAS groups, PRI members, tribal heads, key influential individuals among the local tribes, to motivate community members and create a dialogue against SCD Support ASHAs to train and utilize adolescent peer support groups for awareness generation. Spread importance of pre-marital and pre-conception screening and utility of Sickle Cell Cards Screen children between 6 month to 10 years at anganwadi centers along with PHC-MO/UPHC-MO and RBSK teams.
At School Level	Assist PHC-MO/UPHC-MO and RBSK teams during school visits, parent-teacher meetings and assist in conducting awareness generation, screening and early diagnosis of SCD. Interact with both the students and parents making them aware of the utility of sickle cell cards and the importance of card matching before marriage and conception. Train AB-Health and Wellness ambassadors, to create a dialogue for SCD prevention. Support AB-Health and Wellness ambassadors to organize monthly awareness generation sessions. Distribute iron and folic acid tablets amongst school children. You will administer plain folic acid to those children found to have SCD.

At Facility Level	Ensure display of IEC materials on SCD, SCT, disease transmission, ways of disease prevention, treatment available at the AB-HWC, management and rehabilitation, in local languages Counsel all individuals diagnosed positive with SCD and SCT. Councel adolscents at Adolescent Friendly Health Clinics, to reaching out to adolescents for awareness generation, screening and genetic counselling. Screen pregnant women and counsel for new born screening. Advise on prenatal diagnosis in case both parents are identified as carriers. Continuation on the pregnancy as per the advice of the doctor. Conduct cascade screening of extended family members of identified SCD and SCT cases. Celebrate **World Sickle Cell Day** on **19 June** every year

3.6 Microplanning

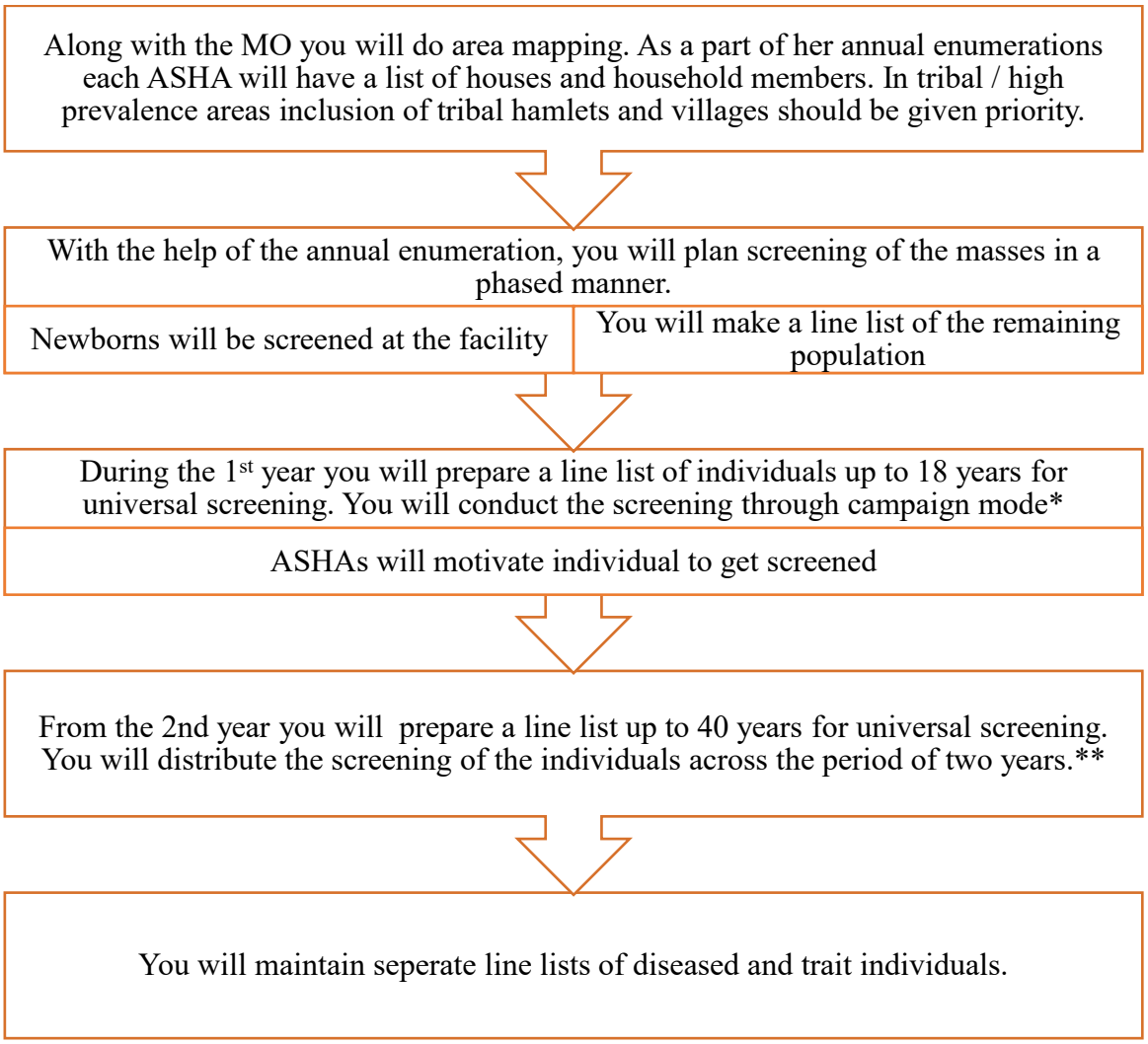
You will support the PHC-MO/UPHC-MO in microplanning the screening activities within the catchment area. In endemic states, blocks will be mapped to identify and grade blocks as per the prevalence. Grading of blocks may be done as below:

Figure 14: Block gradation as per prevalence



Universal screening is to be initially focused on Grade A and B blocks. This may be followed by Grade C blocks. Blocks in Grade D shall require targeted screening approach. Universal screening approach is described in the subsequent flowchart.

Figure 15: Microplanning



* *Population up to 18 years will be covered within the 1st quarter or distributed on a monthly basis but ensure all the screening as early as possible to ensure initiation of comprehensive healthcare for every individual.*

** *Population up to 40 years will be covered on a monthly basis as early as possible to ensure initiation of comprehensive healthcare for every individual.*

Intensive screening will be carried out across the endemic states which will be rolled out for the period of 3 years with the target of 1 crore screening annually each year. For this mapping exercise shall be conducted throughout the endemic states to identify and grade blocks as per the prevalence. The States may screen all newborn upto 18 years of age in first year and incrementally screen all upto 40 years of age.

CHAPTER 4
HOLISTIC MANAGEMENT

4.1 Treatment options available at AB-HWCs

Once the individual is confirmed positive for SCD, the PHC-MO/UPHC-MO will initiate treatment depending on the presenting symptoms. You will maintain a line list of SCD cases and ensure follow-up by motivating the patient for treatment adherence. Treatment of sickle cell disease usually aims at relieving symptoms and avoiding crisis. The range of treatment available at AB-HWCs for individuals suffering from sickle cell disease are as follows:

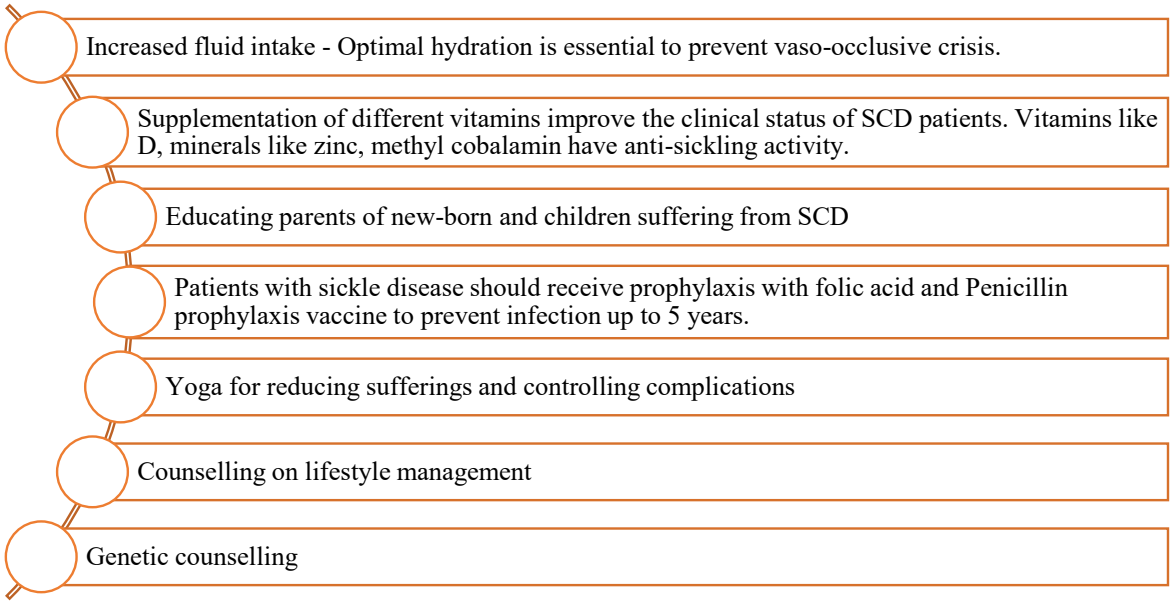
Table 3: Treatment available at AB-HWCs

Preventive	Treatment of Crisis	Rehabilitative	Therapeutic
• Increased fluid intake • Supplementation by different vitamins • Folic Acid prophylaxis • Penicillin prophylaxis to prevent infection up to 5 years. • GoI approved vaccines to prevent infections. • Educating parents of new-born and children suffering from SCD for routine check-up at health facilities • Yoga and wellness • Counselling on lifestyle management	• Crisis episode management which is discussed in details below. • Pain management • Disease modifying therapy ➢ Administration of Hydroxyurea ➢ Referral for blood transfusion • Referral of cases to higher healthcare facilities for ➢ Management of crisis symptoms ➢ Blood transfusion where indicated • Counselling on lifestyle management	• Registration for disability cards • Linkage to Divyangjan (Department of Empowerment of persons with disabilities) for registration and socio-economic support.	• Disease modifying ➢ Administration of hydroxyurea • Referral for blood transfusion, if indicated • Refer patient to higher facilities

4.2 Preventive management

To avoid the crisis episodes or aggravation of symptoms, you will recommend the SCD cases to do the following activities. However, in case of New-born children you will advise the parents to undergo the preventive measures.

Figure16: Activities to prevent aggravation of symptoms



4.2.1 Fluid intake

Dehydration increases the concentration of sickle cell haemoglobin in red blood cells along with risk of vaso-occlusive crisis. You should counsel the patients to maintain optimal hydration through drinking enough fluids, as it is essential to prevent vaso-occlusive crisis.

4.2.2 Nutrition

There is an increased need for vitamins and micronutrients in sickle cell disease. Vitamin D deficiency has been observed in many sickle cell disease patients. Supplementation of different vitamins improves the clinical status of SCD patients. Minerals like zinc, methyl cobalamin, and calcium have anti-sickling activity. You should educate the patients and their family members on inclusion of fruits and vegetables rich in those vitamins and minerals. The following table contains the sources of food items rich in the above-mentioned nutrients.

Table 4: Sources of nutrition

Nutrients	*Source*
Protein	Pulses and legumes, fish, egg, meat, dairy products (milk, curd, cheese), nuts, etc
Carbohydrate of low glycaemic index and rich in fibre	Whole cereals and grains, millets, whole pulses and legumes
Healthy fats used for cooking	Ground nut oil, mustard oil, coconut oil.
Fibre, antioxidants and micronutrients	Locally available seasonal fruits and vegetables of different colours
Vitamin B12	Milk, cheese, curd, paneer, egg, fish etc
Vitamin D	Eggs, paneer, mushroom, milk, fish, cheese, cod liver oil etc
Zinc	Mushrooms, garlic, wheat, watermelon seeds, pumpkin seeds, dark chocolate, cereals, nuts, meat etc.
Calcium	Curd, milk, drumstick leaves, banana, almonds, green leafy vegetables, soya products etc.

The following table highlights the things to do and avoid to attain optimal nutrition:

Table 5: DO's and DONT's in nutrition

DO's	DONT's
Water and other liquids should be given to maintain hydration.	Bakery and confectionary
Frequent meals/ feeds at regular intervals should be offered to achieve the desired goal of nutrition.	Refined cereal and grains
Include natural rich sources of immune nutrients.	Packaged foods, sweetened juices and carbonated beverages.
	Excess fat, sugar and salty food items

Some patients of SCD can have *iron deficiency*- the usual causes are nutritional, parasitic or other etiologies. This may contribute to anaemia and increased infections. Iron deficiency may result in lowering the intracellular haemoglobin concentration and this may ameliorate sickling. Iron deficiency can occur in non-transfusion dependent SCD patients and sickle trait persons. *If iron deficiency is documented it should be treated as in normal persons.*

4.2.3 Educating parents

Educating parents of new-born and children suffering from SCD of developing a habit of bringing their wards to primary healthcare facility at least once a month for follow up. They should also be able to recognize the crisis symptoms to seek help at the earliest.

As a Staff Nurse you should undertake regular check-up of all confirmed patients every 3-6 months. The regular check-up includes:

- Monitoring for fever, jaundice, pallor and spleen size on each health visit
- Monitoring of functions of all vital organs at least once in a year specially when a SCD child grows up to an adult and refer to appropriate centers when indicated.
- Monitoring patients on Hydroxyurea treatment
- Monitoring Haemoglobin levels
- Counselling on diet, stress management and treatment compliance

Sickle cell carriers, usually have mild disease, but may need follow up for regular health maintenance, some will need intervention for fever, pain etc.

4.2.4 Prophylaxis

For preventing complications in persons suffering from SCD the following can be routinely administered. As a Staff Nurse your role will be to follow up on those individuals on the following:

Table 6: Prophylaxis options

Prophylaxis	Dosage
Folic Acid	Daily 5mg
All Vaccines	In accordance with national immunization schedule
Oral Penicillin V potassium	➢ 62.5mg/BD for 1 year ➢ 125mg/day after 1 year until the age of 2 years ➢ 250mg/day till 5 years

4.2.5 Integration of Yoga

Yoga is an integral component of comprehensive primary healthcare provided through Ayushman Bharat Health and Wellness Centres (AB-HWC). Yoga, among other complementary treatments, has the potential to relieve pain in adults and children seeking treatment for acute pain.

4.3 Treatment of crisis

Once the individual is confirmed positive for sickle cell disease, treatment should be initiated. Treatment of sickle cell disease is usually aimed at relieving symptoms and avoiding crisis

4.3.1 Pain management

Pain is the most common symptom of SCD and it may vary in intensity and frequency between patients and can last for few hours or a few weeks. Dehydration, fever, temperature extremes, low oxygen and excessive fatigue are common triggers for pain. As a Staff Nurse you will support the patients presenting with pain symptoms in consultation with the PHC-MO/UPHC-MO.

You should know

Hydroxyurea is contra-indicated during pregnancy and lactation

While on Hydroxyurea therapy the following parameters are to be monitored

- Blood pressure and oxygen saturation at every visit
- CBC count every 2-3 months
- Creatinine and liver function tests every 6 to 12 months
- Urine pregnancy tests as appropriate

4.3.2 Management of crisis

As discussed earlier, it is a very common tendency to develop crisis in cases of SCD. You should be aware of the precipitating factors, symptoms and subsequent treatment. Crisis episodes often require hospitalization, your role as a Staff Nurse will be to identify the crisis and provide palliative care. In complicated cases when patient needs blood transfusion, you will refer the patient to a higher healthcare facility.

If a patient presents with symptoms, you shall conduct the initial evaluation which will include:

- History of pain
- History of prior treatment received prior to arrival at the facility
- Assessment of vital signs: blood pressure, heart rate, respiratory rate, oxygen saturation (administer oxygen if O_2 saturation<90%) and temperature
- Assessment of areas of bone tenderness

4.4 Referral

If the outpatient treatment at PHC-HWC/UPHC-HWC fails to manage pain or patient presents with acute syndrome, sequestration crisis or aplastic crisis then refer the patient to higher center. Danger signs requiring urgent referral includes:

- Chest pain or shortness of breath
- Severe abdominal pain with distension
- Any pain associated with severe pallor

- Joint pain, swelling, redness
- Severe headache or limb weakness.
- Stroke
- Refer severe cases below 2 years of age

4.5 Rehabilitative care

- States extended support to individuals and children suffering from SCD
- Facilities under Rights for Persons with Disability Act, 2016 and amendments

You will make the individuals suffering from SCD aware that they can apply for Disability cards as individuals suffering from SCD are now recognized under Rights of Persons with Disability Act 2016 and amendments. As Sickle Cell Disease is one of the 21 benchmark disabilities under RPWD, any SCD case will be eligible for the following:

- Free education from 6 to 18 years of age.
- 4% reservation in the education and government jobs.
- 5% reservation in higher education
- Other rights under RPWD Act 2016 and amendments

CHAPTER 5
COMMUNITY ADOPTION

5.1 Support through community adoption

Community adoption involves leveraging community support for people with sickle cell disease, as an AB-HWC team member you have to identify, orient, and motivate voluntary individuals and organizations. This initiative shall:

- Provide additional support for patients with sickle cell disease (SCD) to improve treatment outcomes
- Enhance community support in the care of sickle cell disease
- Leverage corporate social responsibility

The community adoption will enhance awareness in the public and active involvement of society about SCD, improve nutritional status of the patients and reduction of out-of-pocket expenditure for the family.

The identified donor can engage to support the consented individual or family of an individual or an entire geography (block/ward/ district) affected by SCD, the types are support are individuals/ families and block/ward/ district.

Table 7: Support from different levels

Individuals/ families	Block/ward/ district
• Nutritional support • Counselling for lifestyle, treatment adherence and stress management • All Vaccines • Mobility support for follow up hospital visits including for penicillin and hydroxyurea prophylaxis • Vocational support	• IEC/BCC activities for community awareness • Organising outreach screening camps • Counselling for lifestyle, treatment adherence and stress management • All Vaccines as per NIS • Vocational support • Genetic Counselling support

5.2 Implementation Plan of Community Adoption:

As a Staff Nurse you will support the PHC-MO/UPHC-MO to plan community adoption and extend support to individuals and families suffering from SCD. Following are the steps in community adoption.

Step 1: Development of integrated web portal and obtaining consent from SCD patients

- Creation of ABHA ID based e-registry for individuals with SCDs
- Integration with centralized application for developing line listing of patients infected with SCDs
- Identification of patients through mass screening campaign and creating line list of all individuals with SCDs
- Mapping of individuals and families with SCDs through frontline workers.

- MO/SN/CHO/MPW/ASHA shall approach the patients directly in person, listed from their area, and inform them about the support available under this intervention. The patient and family shall also be informed that their details shall be made available to the donor.
- Written consent will be obtained from the patient that the enrolment of the patient is his/her informed choice
- For patients who are newly registered in CPHC system, an OTP will be sent to the beneficiary's mobile number and OTP will act as consent for the enrollment.

Step 2: Dissemination of plan

- Use of mass media tools, SMS, digital banners, posters, leaflets, AV testimonials, social media assets, job aids etc.
- Inter-ministerial collaboration to increase public awareness regarding the program.
- Engagement with the mass media at the national and state level.
- Newspaper and TV/radio jingle-based announcements.

Step 3: Donor identification

- Web portal will be developed for self-registration of donors. The page will have provisions to enter the details of the donor, state wise list of districts, blocks, and cities, and the number of existing SCD patients in the block/city. The donor can choose one or more blocks/ urban wards and the intended duration for providing support. They can also enter the type of assistance that they would like to provide for the patients in the area.
- Information about the portal shall be widely disseminated through the use of mass media channels.
- District Health Society (DHS) under the chairmanship of district collector will engage with potential donors across corporates, public sector undertakings, institutions, citizens, elected representatives, etc. DHS Chair shall approve such donations to be finally implemented for the benefit of people with SCD.

Step 4: Service delivery:

- The assistance shall be provided by the identified donor to the patient, as mutually agreed with the district committee on SCD
- The donor and the district committee on SCD shall utilize existing systems or develop new systems to deliver assistance
- The donor should ensure the quality of in-kind assistance provided to the SCD patients

CHAPTER 6
REPORTING AND MONITORING

6.1 Sickle Cell Mobile application

Sickle cell mobile application developed by National Informatics Center shall have data of every person through due generation of ABHA (unique Health ID) after being screened.The indicators pertaining to sickle cell disease intervention at SHC-HWC/UHWC and PHC-HWC/UPHC-HWC shall be updated in the Sickle cell mobile application on a daily basis. These indicators shall be used for reviewing the status of implementation of the sickle disease prevention and care at all levels of care.

6.1.1 Sickle Cell Mobile application

The application has following features:

- Beneficiary registration
- Test details (Solubility, HPLC/Electrophoresis or Point of Care) captured in both online and offline mode
- Integration with ABHA ID

As a Staff Nurse you will register all the individuals to be screened at the facility level and at the outreach camps. The application contains following details for every registered individual:

- Registration: To record the screened person's details.
- Screening Test Details: To record the results of Solubility or POC test.
- HPLC/Electrophoresis Test Details: To record the HPLC/Electrophoresis test details.
- Syncing: To Sync offline (local database) data with the server's data

6.2 Reporting

- ASHAs would conduct population enumeration for detection of probable Sickle Cell Disease and Trait cases. As a Staff Nurse you will ensure monthly reporting of screened and confirmed cases of SCD and SCT, from the facility level.
- You will maintain reports of the following at the PHC-HWC /UPHC-HWC level:

Table 8: Reporting indicators

Total number screened for Sickle Cell Disease
Total number diagnosed with Sickle Cell Trait
Total number diagnosed with Sickle Cell Disease
Total number of persons with Disease registered at the facility
Total number of persons with Disease started on treatment
Total number of persons with Disease under community adoption scheme

- You will ensure that the identified cases are regularly followed up for treatment compliance. You will support the patients requiring any referral to higher facilities.

- You are supposed to update the SCD portal regularly against the total screenings and confirmed cases of SCD and SCT cases.
- You will oversee the tracking of eligible couples through ASHAs, to provide genetic counselling and mobilizing them to the nearest AB-HWCs for SCD screening, prevention and clinical management.
- You will ensure tracking of individuals with known or diagnosed SCD through ASHAs and encourage them to register on Sickle Cell Disease Support Corner, a MoTA's initiative to bridge the gap between patients and health care services in tribal areas.
- Any other responsibility as assigned by the state authorities from time to time.

ANNEXURE A: SOLUBILITY TEST

The solubility test is done to identify altered haemoglobin, either homozygous sickle cell anaemia of heterozygous sickle cell trait.

A.1 Requirements

1. Syringe
2. Blood collection vial lined with anticoagulant
3. Sickle cell buffer reagent – R1
4. Sickle cell powder reagent – R2
5. 2 Vials – one for sample one for control
6. Dropper

A.2 Precautions

1. Store reagent at room temperature
2. Discard reagent on appearance of turbidity, which will not dissolve upon mixing
3. Store the blood sample at 4-8
4. Severe anaemia will cause false negative. Hb concentration should be 7 g/dl or more. Cases of polycythaemia, multiple myeloma, cryoglobulinemia and other dysglobulinemia also have the chances of providing false negative results.

A.3 Preparation of working solution

1. Bring buffer and reagent powder at room temperature before mixing
2. Add one vial of powder reagent (R2) to one bottle of buffer reagent (R1). Cover the cap of R2 bottle and mix vigorously.
3. Record the reconstitution date and expiry date on the vial
4. Store the solubility buffer tightly capped at 2-8
5. Use the buffer within 45 days

A.4 Procedure

1. Prepare the working solution. If already prepared bring to room temperature
2. Collect whole blood in a vial
3. Add 2.0 ml of working solution buffer reagent to two tubes and label sample and control
4. Add 0.02 ml (20µl) of whole blood to the sample vial and plug it. Mix by inversion.
5. Place in the test tube rack for 5 minutes.
6. Read the test in sufficient light after 5 minutes against lines placed in the background

A.5 Inference

1. Positive result (presence of sickled haemoglobin) – If the solution turns turbid and the background lines are not visible

Compare the turbidity of test solution with negative control solution if observed more solution say positive

a. Heterozygous - Red-pink supernatant with a dark red band at the top.

b. Homozygous - Yellowish supernatant with a dark red band at the top.

2. Negative result (no presence of sickled haemoglobin) – If the clear or turbid solution permits the lines to be seen through the tube. Slight greyish matter on top of deep red haemolysate.

3. If in confusion refer the case for electrophoresis

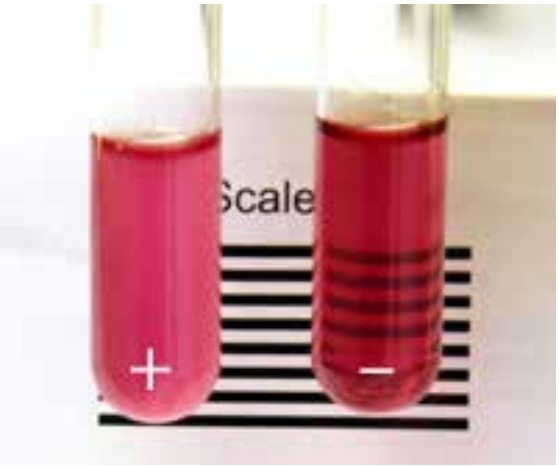


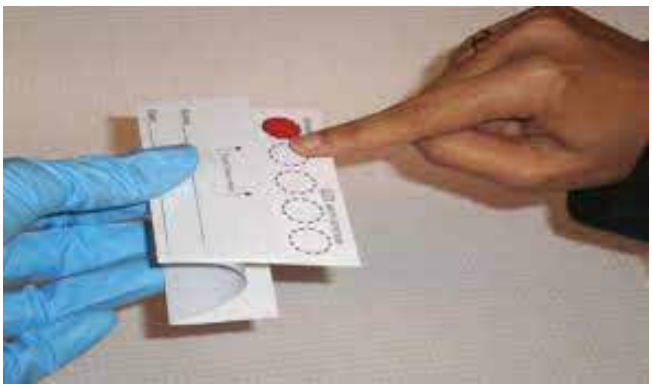
Figure 18: Outcomes of Solubility test

ANNEXURE B. HOW TO TAKE A DRIED BLOOD SPOT SAMPLE

The dried blood spot (DBS) is a simple and quick test that uses the capillary blood from heel collected at the point of testing.

Steps

- Complete the details on the infant request form and blood spot card. Provide as much detail as possible to ensure timely reporting of results.
- Place the infant in a comfortable position with their bare foot. Wash and dry their hands thoroughly and wear appropriate PPE (gloves/apron) during the procedure.
- Clean the area to be used for a blood test with an alcohol wipe and allow to dry
- The best area for the heel prick is the sides of the heel. The sample should not be taken from the back of the heel
- Remove the safety clip and place the lancet in light contact with the heel. This will ensure the incision is not too deep or too shallow. Press the trigger. The blade will create an incision and then retract. Dispose of the device in a sharps box.
- Allow the blood to flow naturally and fill three circles with blood. Fill the circles evenly with one drop of blood per circle. Ensure blood has been absorbed into the filter paper and is visible as a similar size circle at the rear of the card
- If the blood stops flowing, wipe away any congealed blood and gently massage the foot (do not squeeze). Excessive pressure reduces the density of the blood on the sample. If the infant is not bleeding, a second puncture is needed. This should be on the other foot or a different part of the same foot.
- Be careful not to contaminate the sample. Allow the blood spots to air dry for 10 minutes away from direct sunlight or heat. Once completely dry, fold the top of the card over the blood spot and place in the sealed section of the clear plastic bag with the request form in the front pocket



LIST OF ABBREVIATIONS

ABHA	Ayushman Bharat Health Accounts
AB-HWC	Ayushman Bharat - Health and Wellness Centres
AFHC	Adolescent Friendly Health Clinics
ANM	Auxiliary Nurse Midwife
ASHA	Accredited Social Health Activist
BCC	Behaviour Change Communication
CAS	Community Arogya Samiti
CHC	Community Health Centre
CHO	Community Health Officer
CPHC	Comprehensive Primary Healthcare
DEIC	District Early Intervention Centre
DH	District Hospital
DHS	District Health Society
EMRS	Eklavya Model Residential School
Hb	Haemoglobin
IEC	Information Education Communication
JAS	Jan Arogya Samiti
MAS	Mahila Arogya Samiti
MO	Medical Officer
MoHFW	Ministry of Health and Family Welfare
MoTA	Ministry of Tribal Affairs
NIS	National Immunization Schedule
NSAID	Non-steroidal anti-inflammatory drugs
NTAGI	National Technical Advisory Group on Immunization
OPD	Outdoor Patient Department
OTP	One Time Password
PHC-HWC	Primary Health Centre– Health and Wellness Centre
PM-JAY	Pradhan Mantri - Jan Arogya Yojana
PMSMA	Pradhan Mantri Surakshit Matritva Abhiyan
PRI	Panchayat Raj Institute
RBC	Red Blood Cell
RBSK	Rashtriya Bal Swasthya Karyakram
RKSK	Rashtriya Kishore Swasthya Karyakram

RPWD	Rights of Persons with Disability Act
SCD	Sickle Cell Disease
SCT	Sickle Cell Trait
SHC-HWC	Sub Health Centre - Health and Wellness Centre
SDG	Sustainable Development Goal
SN	Staff Nurse
TB	Tuberculosis
TBI	Team Base Incentive
UHC	Universal Health Coverage
UHWC	Urban Health and Wellness Centre
UPHC-HWC	Urban Primary Health Centre – Health and Wellness Centre
VHSNC	Village Health, Sanitation, and Nutrition Committees
VOC	Vaso-Occlusive Crises

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Namaste!

You are a valuable member of the Ayushman Bharat – Health and Wellness Centre (AB-HWC) team committed to delivering quality comprehensive primary healthcare services to the people of the country.
To reach out to community members about the services at AB-HWCs, do connect to the following social media handles:

- https://instagram.com/ayushmanhwcs
- https://twitter.com/AyushmanHWCs
- https://www.facebook.com/AyushmanHWCs
- https://www.youtube.com/c/NHSRC_MoHFW

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