NHSRC

सत्यमेव जयते

Ministry of Health & Family Welfare
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NATIONAL HEALTH MISSION
राष्ट्रीय स्वास्थ्य मिशन

National Sickle Cell Anaemia Elimination Mission 2023

Training Module for Medical Officers






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CONTENT

CHAPTER 1
UNDERSTANDING SICKLE CELL DISEASE

Sickle cell disease is a genetic disorder which is inherited from parents to the child. Normal human Hemoglobin (Hemoglobin A or HbA), also known as adult hemoglobin, (Hemoglobin A1 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 together to produce normal hemoglobin in humans. In sickle cell disease there is substitution of valine for glutamic acid at position 6 of the beta-globin gene. This mutation in the gene leads to formation of abnormal hemoglobin called as sickle hemoglobin. This causes RBC to lose their normal shape and become stiff, sticky curved shape like sickles or crescent moons and lose flexibility. These rigid, sticky cells can get stuck in small blood vessels and cause clogging of blood vessels causing slowing or blocking blood flow and oxygen to parts of the body. This produces pain, results in tissue damage and can lead to serious complications.

lives up to 120 days

Figure 1: Normal red blood cells

Figure 2: Sickle cell

live up to 10-20 days

1.1 TYPES OF SICKLE CELL

HbSS: People who have HbSS inherit sickle cell genes ("S"), from both parents and have sickle cell disease(SCD)

Sickle Cell Trait (SCT): People who have SCT inherit one sickle cell gene ("S") from one parent and one normal gene ("A") from the other parent. 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 to their children. Exposure to extreme stressful conditions, can lead to crisis which may require repeat check-ups

HbS beta thalassemia: People who have this form of Sickle Cell disease inherit one sickle cell gene ("S") from one parent and one gene for beta thalassemia, a type of anaemia, from the other parent. Those with HbS beta thalassemia usually have a severe form of SCD

1.2 I INHERITANCE OF SICKLE CELL

A pictorial depiction of different combinations of parents' genetic status and the probability of the children getting affected with sickle cell disease is as below:

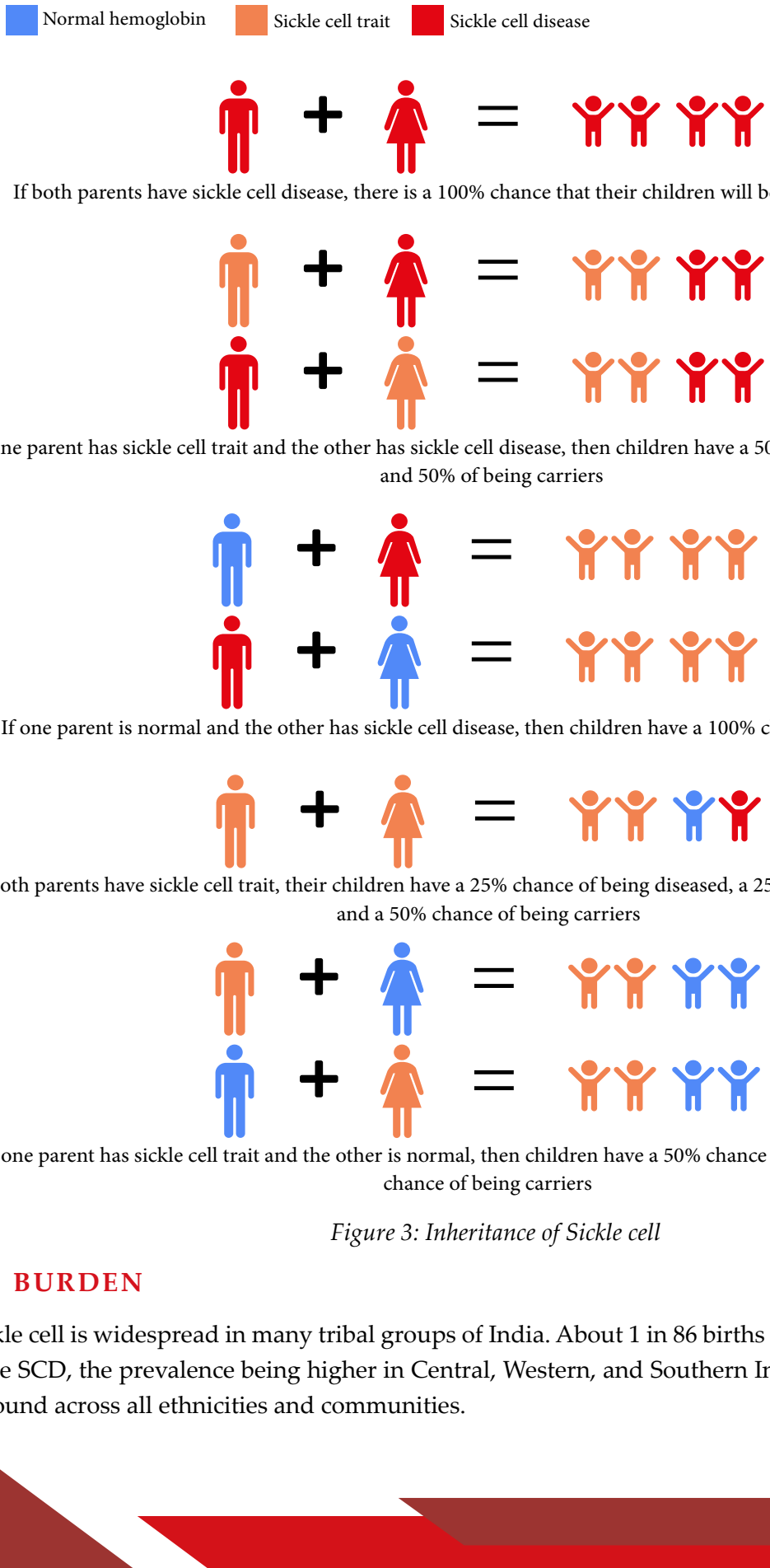


Figure 3: Inheritance of Sickle cell

1.3 BURDEN

Sickle cell 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.

States with the prevalence of Sickle Cell Disorder include- Gujarat, Rajasthan, Uttarakhand, Maharashtra, Bihar, Jharkhand, Madhya Pradesh, Chhattisgarh, Odisha, West Bengal, Tamil Nadu, Telangana, Andhra Pradesh, Karnataka, Kerala, Uttar Pradesh & Assam.

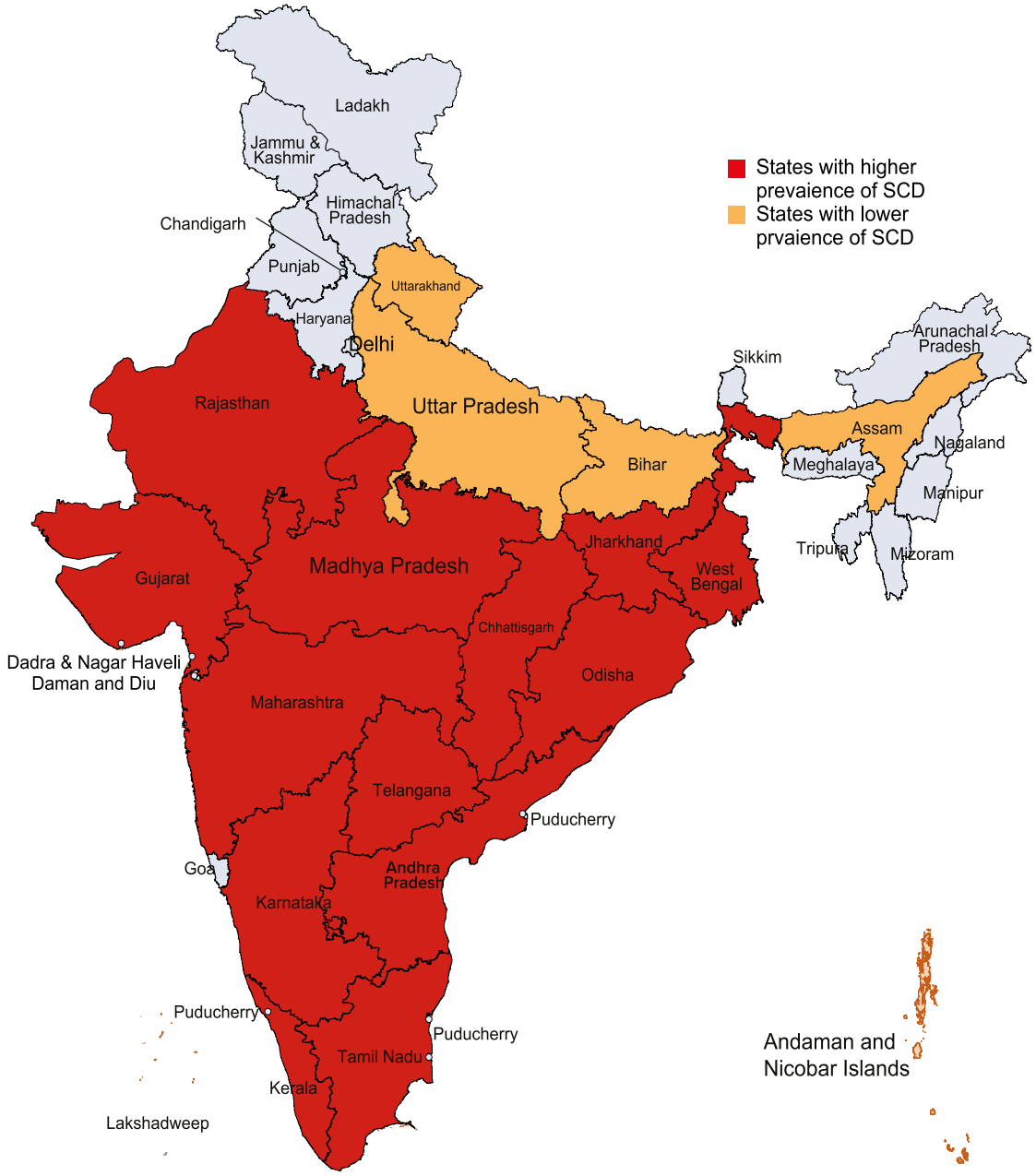


Figure 4:States with prevalence of Sickle cell

1.4 SOCIO-ECONOMIC IMPLICATION

As per the global burden disease 2019 report ,sickle cell disease accounted 48.7 million disability adjusted life years (DALYS).[1] SCD has a significant macroeconomic effect on individuals as well as on their family. Direct costs are those that are met by the healthcare system and those these include

[1](https://www.healthdata.org/results/gbd_summaries/2019/sickle-cell-disorders-level-4-cause)

- 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.

Suffering from SCD has indirect effect on the socio-economic condition

- Loss of productivity because of parents' missing days of work to look after their children.
- Future loss to the economy as sickle cell disease deaths mainly occur in children and young adults causing early deaths that erode 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

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 Anaemia Elemination 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 and quality care 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 levels 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 OF SICKLE CELL DISEASE ELIMINATION PROGRAM

The Sickle cell disease elimination program is a part of National Health Mission and focuses on universal population-based screening, prevention, and management of sickle cell disease in high prevalence 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 minimizing the duplication of efforts, for example, established platform of 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, Auxiliary Nurse Midwifes, Multi-purpose workers (M/F), and Accredited Social Health Activists (ASHA) 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, EARLY IDENTIFICATION AND DIAGNOSIS

One major intervention for achieving the goal of a sickle cell-free nation is screening and early diagnosis.

3.1 SCREENING APPROACH

- **Option 1:** One step approach-Point of Care test (POC) 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. This GoI approved electrophoresis based POC can be used in small setting where lab technicians are placed. The test requires pipette with five disposable tips and slide for mixing of buffer. The five tips and slide per test needs proper disposal as biomedical waste.
- **Option 2:** Two-step approach- Mass screening / Initial screening using Solubility test (Annexure1). If found positive for Solubility test in field setting, then confirmation using Point of Care confirmatory test or HPLC / Electrophoresis at higher centers.

3.2 TARGET GROUPS

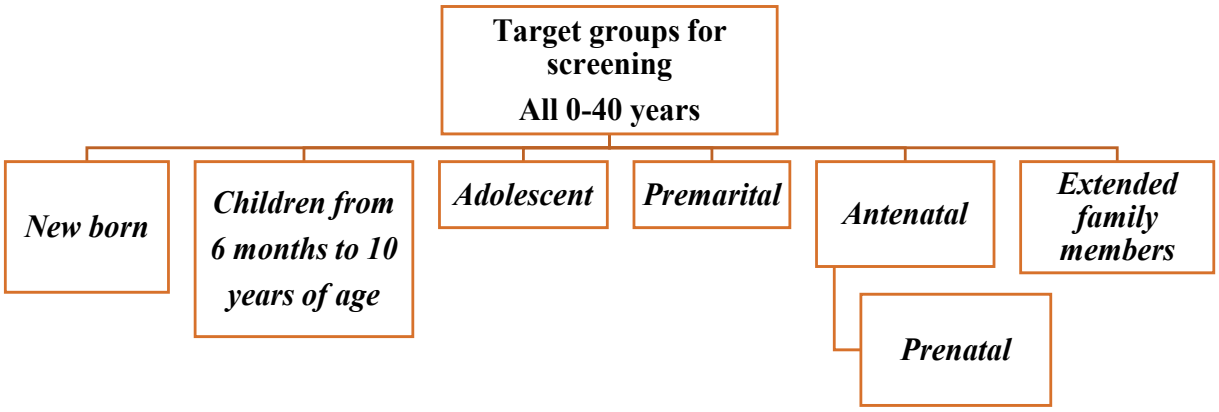


Figure 5: Target groups for screening

3.3 TYPES OF SCREENING

3.3.1 Mass screening: This will be organized by primary health care team at PHC-HWC, UPHC-HWC, SHC-HWCs and UHWCs. All the eligible individuals will be mobilized for screening by the ASHA and MPW(M/F) in collaboration with Jan Arogya Samitis (JAS) and Village Health Sanitation and Nutrition Committees (VHSNC) or Mahila Arogya Samiti (MAS),Community Arogya Samiti (CAS). Solubility test would be used for mass screening of population.

As a Medical Officer you shall :

- **Ensure** every new-born born at the facility is screened for Sickle cell
- **Conduct outreach screening camps on a monthly basis**-focus should be on screening of individuals from zero up to 18 years of age in the first year and then up to 40 years of age in the next year. Other community based platforms like VHSNC, SHG should be engaged.
- **Ensure microplanning based on the area mapping exercise by ASHA-** Total number of houses and household members shall be enlisted by ASHA as a part of her annual enumerations. Line listing of diseased and trait individuals shall be maintained separately.
- **Ensure that** ABHA ID linkage and Sickle cell card are provided to the screened individuals.
- **Counsel** all those who are being screened on sickle cell disease prevention and treatment modalities.
- **Ensure that** individuals identified positive are referred to the nearest secondary care facility (CHC/DH) in case of complication/emergency.

3.3.2 Opportunistic screening: This will be done at the facility level. Solubility test would be adopted for screening. Those found reactive will be further tested by Point of Care test. Treatment will be initiated on confirmation. MO may recommend the individual to higher facility for confirmation through high performance liquid chromatography (HPLC)/electrophoresis which is a confirmatory test with high sensitivity and specificity.

As a Medical Officer you shall :

- screen for people attending outpatient services at the PHC-HWC,UPHC and UHWC level
- prioritize individuals belonging to the following categories
    - Antenatal screening (early 1st trimester) of all pregnant women for sickle cell carrier status. If any pregnant woman is found to be a sickle cell carrier, then her husband too would be tested for carrier status
    - Refer pregnant women to higher facilities for prenatal testing who are found positive for sickle cell trait or disease
    - Universal screening of all new-born and people below 40 years of age
    - Cascade screening of extended family members of carriers and sickle cell positive people
- ensure that each individual being screened shall be registered with a ABHA ID and provided with a Sickle cell card in case the person is diseased or have trait
- follow- up of the confirmation of disease or trait status
- ensure that individuals with confirmed sickle cell disease diagnosis shall be registered in Sickle cell portal and are provided with sickle cell card. The card will include the status of the screening individual (Normal, Carrier, Diseased).

3.3.3 Intensive screening: This 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.

Grading of blocks are as below:

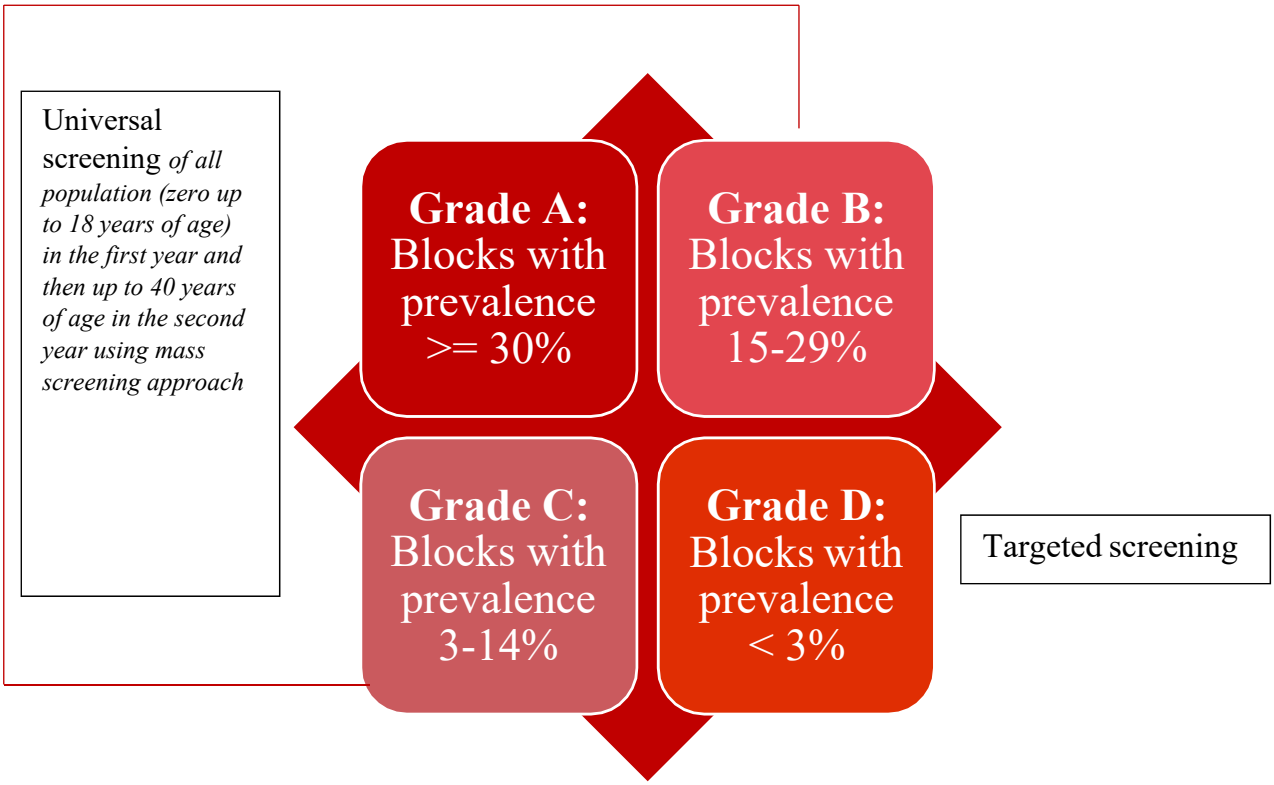


Figure 6.Grading of Blocks

As a Medical Officer you shall :

- **Ensure** that mapping exercise is conducted to identify and grade blocks based on prevalence
- **Microplan** based on mapping exercise to initiate universal screening. This will include deploying of primary health care team, logistics management and identification of target population based disease prevalence.
- **Ensure** universal screening is initially focused on Grade A and B blocks followed by Grade C blocks.
- **Ensure** targeted screening for groups mentioned in figure 6 or blocks in Grade D

3.4 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 will be color coded separately for male (blue) and female (pink). Based on the card's status, the individual will receive treatment and counselling services. The Sickle cell cards will be extensively used for the purpose of pre-marital and pre-conceptional counselling by matching the cards of prospective matches. Matching of the cards will show the chances of their children being born with SCD or SCT.

CHAPTER 4
HOLISTIC MANAGEMENT AND CONTINUUM OF CARE

The range of treatment available at AB-HWCs for individuals suffering from sickle cell disease are as follows:

Table 1: Management of Sickle cell

Preventive	Treatment of Crisis	Rehabilitative	Therapeutic
• Increased fluid intake • Supplementation by different vitamins • Folic Acid prophylaxis • Penicillin prophylaxis • 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 • Referral of cases to higher healthcare facilities for management of crisis symptoms • Referral for blood transfusion where indicated • Counselling on lifestyle management	• Registration for disability cards • Linkage with Divyangjan (The Department of Empowerment of Persons with Disabilities)	• Disease modifying therapy • Administration of Hydroxyurea • Referral for blood transfusion, if indicated • Refer patient to higher facilities

4.1 PREVENTIVE MANAGEMENT

4.1.1 Genetic Counselling

According to WHO (2016), genetic counselling is the most cost-effective intervention to reduce the incidence of hemoglobin disorders, including SCD. Genetic counselling is a communication process in which the counsellor is expected to take into consideration the emotional aspects of the individual as well as ensure clients gain a thorough understanding of the condition. Furthermore, it is beneficial for premarital decisions among would-be couples, particularly carriers, to understand the risks of possible disorder transmission to their children. Genetic counselling includes:

a) Pre-marital counselling

As a Medical officer you shall counsel the individuals about the risk of having affected children, if they marry an individual with either trait or disease with the help of sickle cell cards.

The card has details such as gender, test report (Sickle cell disease/Sickle cell carrier/ Normal) on the front side of the card. 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 disease or trait in the child. The following are the matching possibilities:

- 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

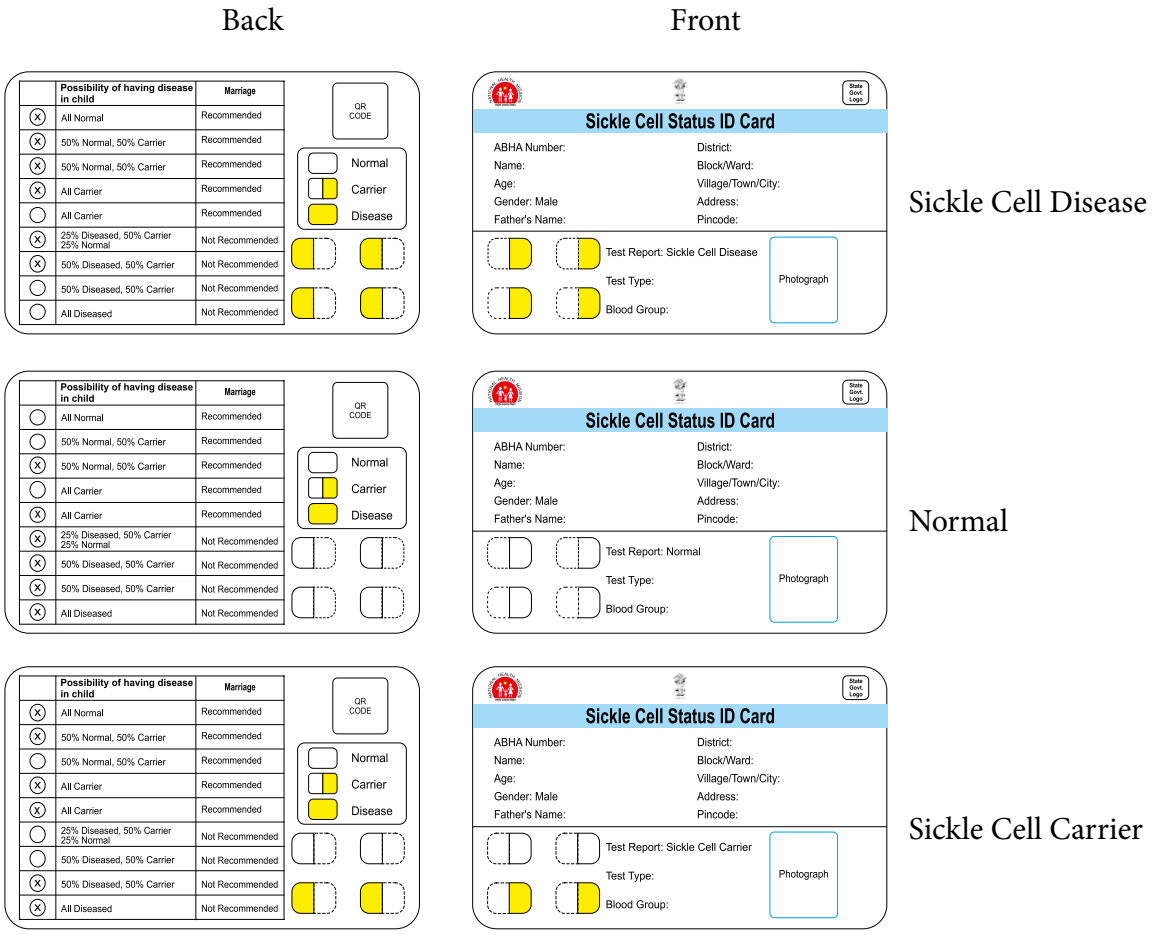


Figure 7: Sickle cell cards for males

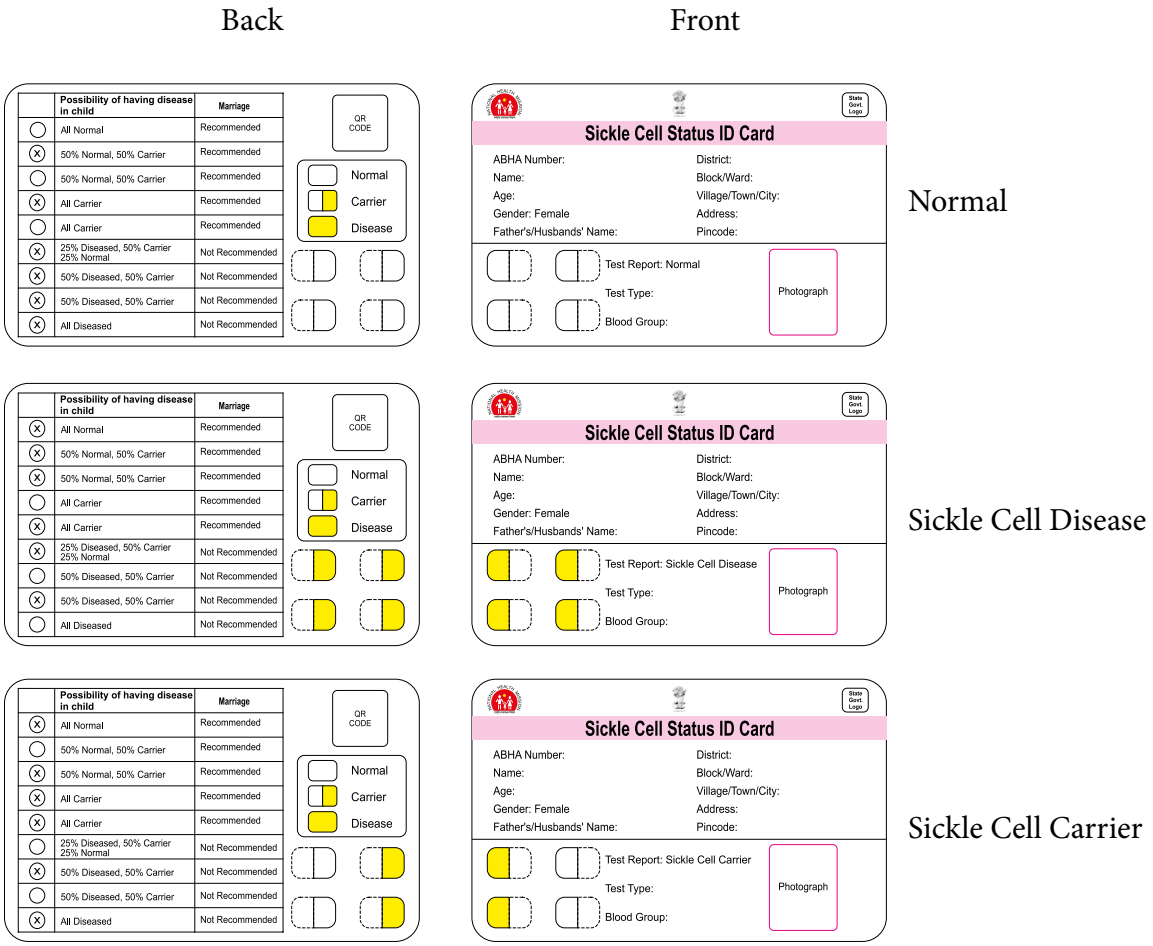


Figure 8: Sickle cell cards for females

b) Pre-conceptional counselling

Pregnancy with Sickle cell anaemia is of high risk category with increased maternal complications such as anaemia, vaso-occlusive (VOC) crises, acute chest syndrome (ACS), jaundice, preeclampsia, urinary tract infections and mortality. A woman with SCD whose partner is a carrier will have a risk of up to 50% in each pregnancy of having a child with a sickling disorder. As a medical officer your role will be to

- Counsel women with SCD/SCT to get her partner screen prior to the initiation of pregnancy
- Counsel for reproductive options, and refer for prenatal diagnosis for high-risk couples. Only two combinations warrant referral to a tertiary care centre for pre natal testing and diagnosis, as shown below:

Prenatal diagnosis required	Prenatal diagnosis not required
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 (to know the carrier status of the child)	If two individuals having sickle cell disease marry, there is a 100% chance that their children will be born with SCD. They should be counselled and helped in an informed decision making regarding continuation of the pregnancy
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 (to know the carrier status of the child)	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 a sickle cell disease trait and a normal individual marry, their children have 50% chance of being normal and 50% chance of being carriers

4.1.2 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 a thorough check up. They should be educated to recognize the crisis symptoms to bring their wards to the primary healthcare facility.

4.1.3 Prevention of Sickle Cell Crisis

Management of sickle cell disease is largely driven by medication. However, nutrition also plays an important role as sickle cell patients need more energy intake than normal person because of elevated energy expenditure related to physiological adaptations. As a medical officer you can advise on following:

- *4.1.3a Fluid intake*

  Dehydration increases the concentration of sickle cell hemoglobin in red blood cells along with risk of vaso-occlusive crisis. Maintaining of optimal hydration through drinking enough fluids is essential to prevent vaso-occlusive crisis.

- *4.1.3b Vitamins and micronutrients*

  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, calcium, methyl cobalamin, have anti-sickling effect. Patient education on inclusion of fruits and vegetables rich in these vitamins and minerals should be done.

- *4.1.3c Nutrition support to the SCD patients*

  - Good Nutrition with extra calories, and anti-oxidants from food are important and would help to reduce the growth inhibition seen in SCD patients. The resting metabolic rates (RMR) in SCD children is higher than normal children. Protein metabolism consumes approximately 30% of energy at rest, and recent data indicate that the high resting energy expenditure (REE) in HbSS is determined primarily by energy needs for cardiac compensation and increased protein metabolism.

- 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 hemoglobin 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 with iron folic acid supplementation.

Following can be recommended:

- Prefer good quality of carbohydrate of low glycemic index and rich in fiber to maintain a healthy gut and immunity e.g. whole cereals and grains, millets, whole pulses and legumes.
- Include vegetable and animal sources of protein in the diet to achieve high protein intake, such as nuts, milk, pulses and legumes or egg, fish, meat.
- Healthy fats should be used for cooking e.g. fats from plant origin and nuts &seed oils e.g. Ground nut oil, mustard oil, coconut oil.
- Include locally available seasonal fruits and vegetables of different colors in the diet to meet required fibre, antioxidants and micronutrients.
- Water and other liquids should be given to maintain hydration.
- Frequent meals/ feeds at regular intervals should be offered to achieve the desired goal of nutrition.
- Include natural rich sources of immune nutrients.

Restrict foods which lower immunity such as

- Bakery and confectionary
- Refined cereal and grains
- Packaged foods, sweetened juices and carbonated beverages.
- Excess fat, sugar and salty food items

- *4.1.3d Lifestyle modification*

This includes avoidance of stress, exertion, avoidance of dehydration and extreme climatic condition to minimize acute complications

4.2 Management of persons with sickle cell disease

You should be aware of the clinical presentation of sickle cell disease for curative management.

4.2.1 Clinical presentation

A suspected sickle cell disease (SCD) patient will most likely present with a range of clinical manifestations such as pallor, pain, fever, lethargy, and jaundice. Sickle cell disease can cause both acute and chronic complications.

Acute complications include:

- Infections
- Severe anaemia (can be caused by splenic sequestration, aplastic crisis, or hyper- homolysis).
- Vaso-occlusive phenomena manifesting as pain, stroke, acute chest syndrome, renal infarction or medication toxicity, dactylitis or bone infarction, and myocardial infarction.

- Pregnancy complications, such as spontaneous abortions, stillbirths, and IUGR
- Priapism and venous thromboembolism are also common.
- Stroke

Chronic manifestations include :

- Chronic pain, anaemia, neurologic deficits or seizure disorder
- Pulmonary conditions such as pulmonary hypertension
- Renal impairment
- Osteoporosis and bone infarction
- Cardiomyopathy with diastolic dysfunction, hepatotoxicity and pigmented gallstones
- Chronic leg ulcers, and proliferative retinopathy

4.2.2 Sickle Cell Crisis

- **Vaso-occlusive crisis:** A vaso-occlusive crisis occurs when the microcirculation is obstructed by sickled red blood cells resulting in ischemic injury. The major complaint is pain, usually affecting bones such as femur, tibia and lower vertebrae. Alternatively, vaso-occlusion may present as dactylitis, hand and foot syndrome (painful and swollen hands and/ or feet), or an acute abdomen. The spleen may undergo auto-infarction and is often not palpable beyond 6 year of age. Involvement of the kidney results in papillary necrosis leading to inability to concentrate urine (isosthenuria). Other presentations include acute chest syndrome, retinal hemorrhages, priapism, jaundice, avascular necrosis of the femoral head and cerebrovascular accidents.
- **Acute chest syndrome:** This is a type of Vaso-occlusive crisis that affects the lung and presents with chest pain, cough, tachypnoea, dyspnea, 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-limiting and may follow viral infections of which parvovirus B19 is the most commonly implicated.

4.2.3 Relieving Symptoms

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. 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, extreme temperature, low oxygen and excessive fatigue are common triggers for pain.

As a Medical officer you shall:

- **Manage** mild to moderate pain through acetaminophen unless there is any contraindication.
- Ibuprofen in dose of 10mg/kg/8hours can be given but used with caution in patients at high risk of acute chest syndrome. The use of NSAIDs should be avoided during pregnancy
- Acetaminophen in dose ranging from 12.5mg/kg/dose IV every 4 hours to 15 mg/kg/dose IV every 6 hours. IV Acetaminophen reduces pain from vaso occlusive crisis in children with sickle cell disease.

- **Consider** Hydroxyurea for patients experiencing repeated episodes of acute chest syndrome or with more than three crises per year requiring hospitalization. It has been proven to decrease complications, such as - pain crisis, acute chest syndrome and strokes. Hydroxyurea can be administered to severe SCD in consultation with pediatrician/hematologist cases above 2 years. Children below 2 years cases should be referred to higher centers. The starting dose of Hydroxyurea is 10 mg/kg/ day with other supportive care and followed for one year or more. It is contraindicated during pregnancy and lactation.
- Since patients with SCD are susceptible to both viral and bacterial infections, you should consider any fever or infection as a medical emergency.

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

Figure 9: Monitoring parameters when patient is on Hydroxyurea

4.2.4 Management of Crisis

Sickle crisis refers to worsening of symptoms of SCD over a short period of time. This may require urgent referral for hospitalization. As a medical officer you should be aware of the precipitating factors for crisis, symptoms and management at AB-HWC level.

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 hospital
- Assessment of vital signs: blood pressure, heart rate, respiratory rate, oxygen saturation (administer oxygen if O2 saturation<90%) and temperature
- Assessment of areas of bone tenderness

The details of management of crisis is given below:

Table 2: Crisis presentation and management

Crisis	Precipitating factors	Symptoms	Treatment
Vaso-occlusive crisis	Exercise, infection, dehydration, psychological stress, exposure to extremes of temperature	Pain in joints like knee, shoulder and elbow, small joints in children (3-10 years) and in back and chest in adults.	Pain relief medication Oral Hydration Reassurance Anticoagulants can be started in in consultation with hematologists

Acute chest syndrome	Infection and over hydration	Wheezing, cough, increased work of breathing, and fever in children and chest pain, pain in the arms and legs, shortness of breath in adults Oxygen saturation below 95-92 X-ray showing new infiltrates	Pain control, appropriate intravenous (IV) fluids, antibiotics, supplemental oxygen. Referral for • Appropriate blood transfusions • Ventilatory support if needed
Sequestration crisis	Recurrent episodes	Sudden pallor, Weakness, with sudden increase in the size of spleen, severe anemia and shock	Referral for • Blood transfusion • Splenectomy - post 3 splenomegaly crisis episodes Immunization and Prophylactic penicillin for 3 months as protective measure following splenectomy
Aplastic crisis	Infection (parvovirus B19)	severe anemia	Referral for blood transfusion

4.3 PROPHYLACTIC MANAGEMENT

- Folic acid should be given once the diagnosis of SCD is made to prevent folic acid deficiency due to chronic homolysis and also for those patients who are on Hydroxyurea.
- You should administer routine immunization to a newborn as per national immunization schedule.
- Children with SCD are significantly more likely to develop invasive pneumococcal disease; however, daily antibiotic prophylaxis until the age of 5 years significantly reduces the risk. Pneumococcal bacteremia is reduced by daily penicillin prophylaxis. In infants with sickle cell disease, splenic dysfunction begins as early as 3 months. All of these children should be started on prophylactic Penicillin treatment as early as 2-3 months of age. This prophylaxis should be continued for at least five years. If the child had a splenectomy or had pneumococcal infection, prophylactic penicillin should be continued beyond five years. The dosage of penicillin is as follows:
    - 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
- As a medical officer your role will be to follow up on those individuals on penicillin prophylaxis.

4.4 REFERRAL

If the outpatient treatment at PHC-HWC, UPHC-HWC, and UHWC fails to control 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
- In case of children below 2 years with severe illness

4.5 FOLLOW-UP

As a medical officer 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
- Monitoring functions of all vital organs and systems at least once in a year specially when a SCD child grows up to an adult and refer to appropriate centers.
- 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.

4.6 REHABILITATIVE CARE

You shall make the individuals with SCD aware of the following:

- States have extended support to individuals and children suffering from SCD
- that they can apply for Disability cards as individuals with SCD are now recognized under Rights of Persons with Disability Act 2016.
- Facilities under Rights for Persons with Disability Act, 2016 and amendments.
- As Sickle Cell Disease is one of 21 benchmark disabilities under RPWD, any individual with SCD 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

4.7 INTEGRATING YOGA IN SICKLE CELL DISEASE CARE

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, according to the American Society of Hematology Guideline 2020 panel. However, they did not, recommend yoga as a treatment modality for chronic pain.

CHAPTER 5
HEALTH PROMOTION FOR SICKLE CELL DISEASE

Health promotion is the process of empowering people to gain more control and improve their health. One of the most important health promotion interventions is health education, which is the most cost-effective way of preventing disease with little or no medical intervention if people are adequately informed and take the necessary precautions on time. In case of sickle cell disease awareness on the understanding and management of the disease is important for patient and educating their family. This can be strengthened through the following :

5.1 ROLE OF COMMUNITY PLATFORMS

Village Health Sanitation and Nutrition Committee (VHSNC) meetings, Village/Urban Health Sanitation and Nutrition Days (VHSND/ UHND) meetings at Anganwadi, Jan Arogya Samitis (JAS) in AB-HWC, Mahila Arogya Samiti (MAS), Community Arogya Samiti (CAS), Arogya Sabha, Self-Help Groups (SHG), youth clubs, parent-teachers meetings in schools, etc., are an important platforms to make people aware on the importance of sickle cell disease. Your role as Medical Officer would be to:

- Plan and undertake monthly health promotion activities/campaigns to improve community awareness and
- To sensitize people on the importance of sickle cell disease and screening service available at AB-HWCs
- Ensure that PRI members, tribal head, key influential individuals among the local tribal, especially women as champions and other community shall be engaged to spread awareness and motivate community members to visit AB-HWCs, attend outreach camps and utilize the screening and counselling services at the nearest facility
- Ensure that locally relevant awareness modalities such as street plays, miking, wall writings and paintings, quizzes, etc, shall be undertaken to raise community awareness on sickle cell disease and the national mission
- Educate school teachers and anganwadi workers about the special needs of children with sickle cell disease through conducting health talks and counselling session.
- Facilitate training of the Ayushman Bharat -Health and Wellness ambassadors to enable them to transact health promotion and disease prevention information in form of interesting activities which would be either classroom based or as an outreach activity.
- Utilize the existing mechanism under School Health Programme to ensure SCD related activities across schools.
- Ensure all SCD related interventions at school level through Eklavya Model Residential School (EMRS), the flagship initiatives of GoI.
- Leverage the platform of Health Mela for all SCD screening and counselling services

5.2 ROLE OF PATIENT SUPPORT SYSTEM

Patient Support Group formation helps in treatment adherence by reducing social stigmas and increasing disease acceptance. With the increasing prevalence of chronic illnesses, the concept of a patient support group may really help you in health promotion for populations suffering from similar illnesses and addressing their common concerns. For individuals with sickle cell disease, as a medical officer you can ensure that:

- Patient support groups are created after area mapping based on disease burden
- Micro-plan the meeting with members once the group is formed, with the help of CHO and ASHA
- At the meetings, people with disease share their experiences, which may aid them in identifying complications early and attaining support for treatment compliance
- Ensure that every session has discussion over disease management, early diagnosis and treatment adherence.

5.3 COMMUNITY ADOPTION

Community adoption involves leveraging community support for people with sickle cell disease, as a Medical Officer 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 reduce 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.

5.4 IMPLEMENTATION PLAN OF 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/UT 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
MONITORING, SUPERVISION AND REPORTING

6.1 MONITORING

Sickle Cell Mobile application developed by National Informatics Center shall have data of every person after 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

The application contains following details:

- 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

As a medical officer you should

- Register your mobile number through concerned District User / State User / State Admin
- Download mobile application through https://sickle.nhm.gov.in/home or https://sickle.nhm.gov.in/home/app_download_counts
- Login using mobile number and generate MPIN
- Register the screened people through ABHA ID

6.2 SUPERVISION AND REPORTING

As a Medical Officer you shall:

- Ensure monthly reporting from facility level includes data pertaining to the screening, patients identified and on treatment, persons lost to follow up (reporting format)
- Ensure monthly data is uploaded in the portal
- Provide technical support to the CHO/Staff Nurse/Pharmacist/Lab Technician/MPW(M/F)/ ASHA for appropriate maintenance of records and reports on screening, treatment, counselling, referral and follow up

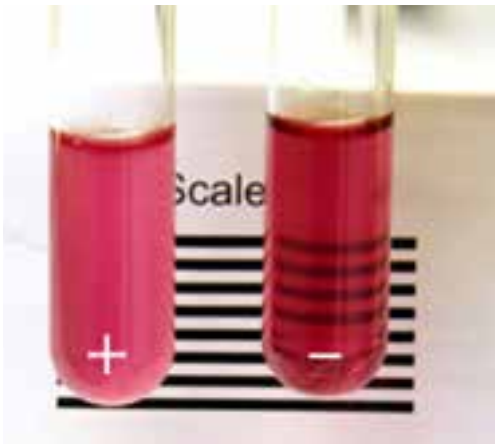
- Ensure availability of logistics, diagnostics and consumables as required for the camps for the PHC-HWC/UPHC-HWC as well as catchment SHC-HWC/UHWC
- Train and mentor the team at SHC-HWC/UHWC level–on all aspects of SCD prevention, control, counselling and management
- Monitor the conduction of community/village level meetings such as VHSNCs or campaigns conducted
- Facilitate of mapping of patients for community adoption
- Monitor suggested set of 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

- Follow -up identified cases for treatment compliance. Any referral to higher facilities should be supported through the AB-HWCs.
- Track eligible couples, to provide genetic counselling and mobilizing them to the nearest Ayushman Bharat Health and Wellness Centre (AB-HWC) for SCD screening, prevention and clinical management.
- Track individuals with known or diagnosed SCD should be encouraged to register on Sickle Cell Disease Support Corner, a Ministry of Tribal Affairs initiative to bridge the gap between patients and health care services in tribal area.

Annexure 1. Solubility test

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



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

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 4-8C
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.

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

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

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

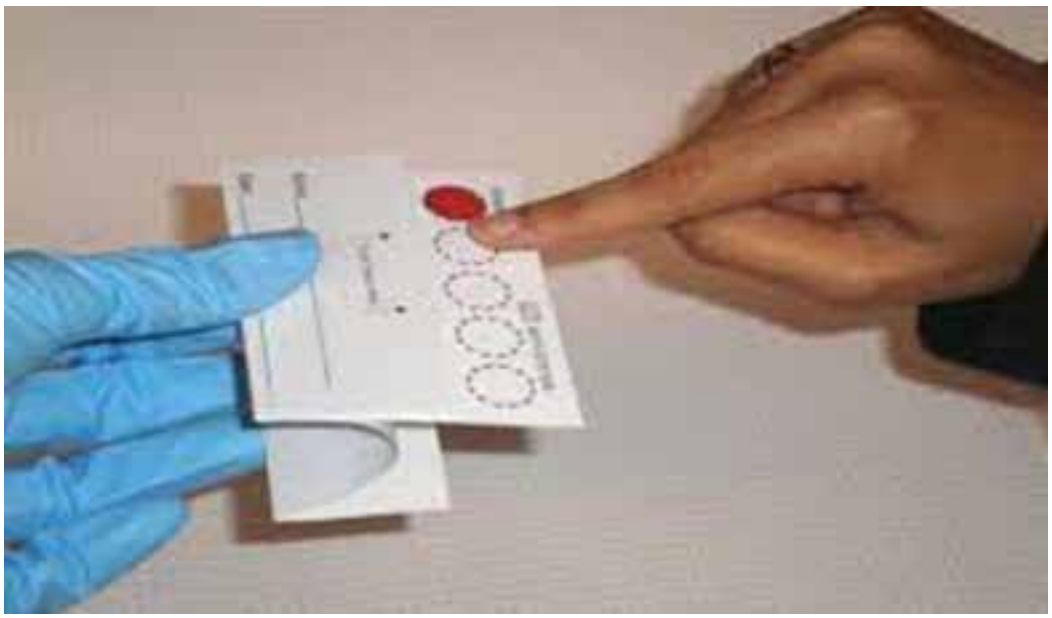
   a. Heterozygous - Red-pink supernatant with a dark red band at the top.
   b. Homozygous - Yellowish supernatant with a dark red band at the top.
2. Negative result (no presence of sickled haemoglobin) – If the clear or turbid solution permits the lines to be seen through the tube. Slight greyish matter on top of deep red hemolysate.

Annexure 2. 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



Annexure 3. Abbreviation

ABHA ID	Ayushman Bharat Health Account ID
ACS	Acute chest syndrome
ANM	Auxiliary Nurse and Midwife
ASHA	Accredited Social Health Activist
AV	Audio visual
CHC	Community Health Center
CHO	Community Health Officer
CPHC	Comprehensive Primary Health Care
DALY	Daily adjusted life years
DH	District Hospital
DHS	District Health System
EMRS	Eklavya Model Residential School
GoI	Government of India
Hb	Hemoglobin
HPLC	High Performance Liquid Chromatography
IUGR	Intrauterine growth retardation
JAS	Jan Arogya Samiti
MAS	Mahila Arogya Samiti
MO	Medical Officer
MoHFW	Ministry of Health and Family Welfare
MoTA	Ministry of Tribal Affairs
NHM	National Health Mission
NIS	National Immunization Schedule
NSAID	Non-steroidal anti-inflammatory drugs
NTAGI	National Technical Advisory Group on Immunization
PHC-HWC	Urban Primary Healthcare center-Health and wellness center
PRI	Panchayati Raj Institutions
RBC	Red blood Cell
RBSK	Rashtriya Bal Swasthya Karyakram
RPWD	Rights of Persons with Disabilities
SCD	Sickle cell disease
SCT	Sickle cell trait
SHG	Self Help Group
SMS	Short message service
UPHC-HWC	Urban Primary Healthcare center-Health and wellness center
VHND	Village Health Nutrition Day
VHSNC	Village Health Sanitation and Nutrition Committees
VOC	Vaso-Occlusive crisis
WHO	World Health Organization

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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

National Health Systems Resource Centre

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