Clinical Features of Sickle Cell, Thalassemia Disease and Role of Counsellors

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What is sickle cell disease?

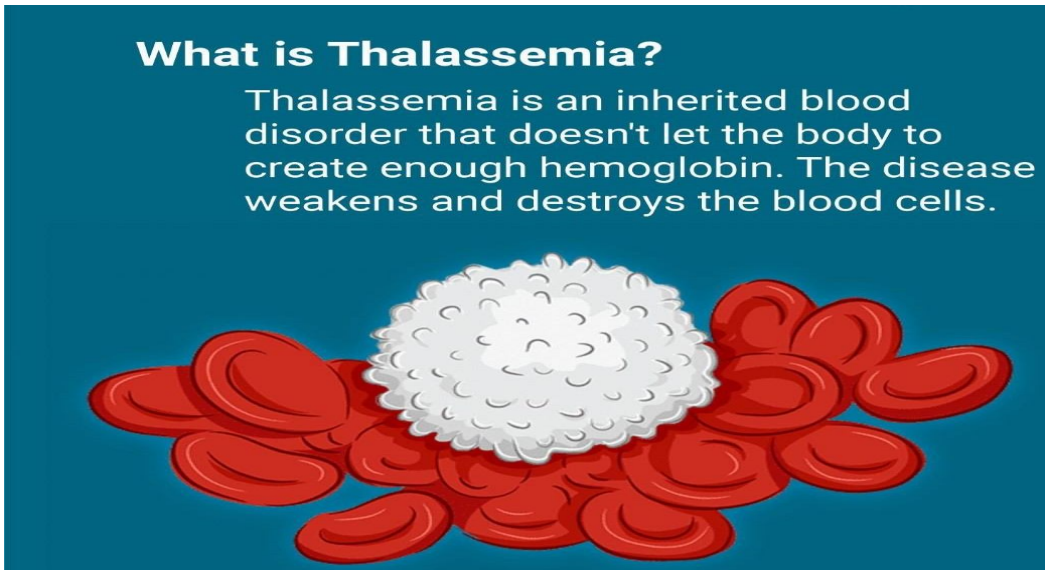
- Sickle cell anaemia is a genetic condition where the haemoglobin molecule inside red blood cells is abnormal.
- The red blood cells carrying sickle-haemoglobin become sticky and deformed (C shaped)
- This can block small blood vessels causing severe pain, and organ damage including stroke.
- These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

Normal RBC and Sickle RBC

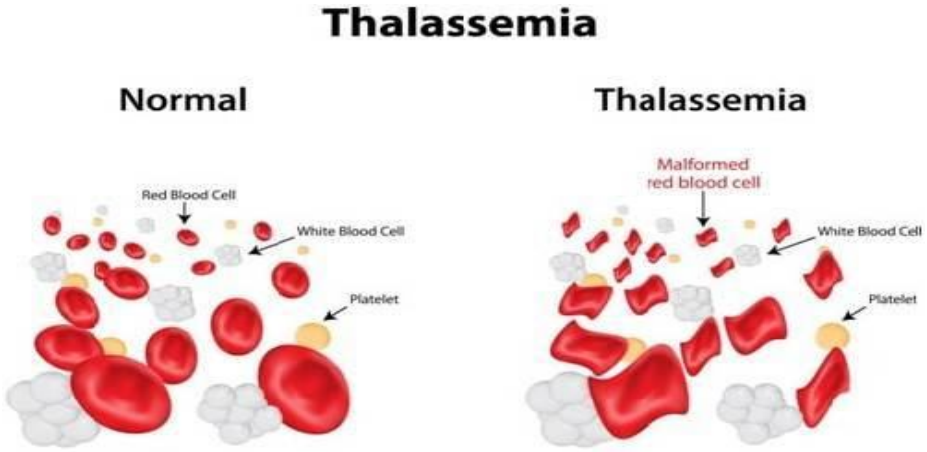
Clinical Features of Sickle Cell Disease

- Anemia. Sickle cells break apart easily and die.
- Episodes of pain. Periodic episodes of extreme pain, called pain crises, are a major symptom of sickle cell anemia.
- Swelling of hands and feet.
- Frequent infections.
- Delayed growth or puberty.
- Vision problems.

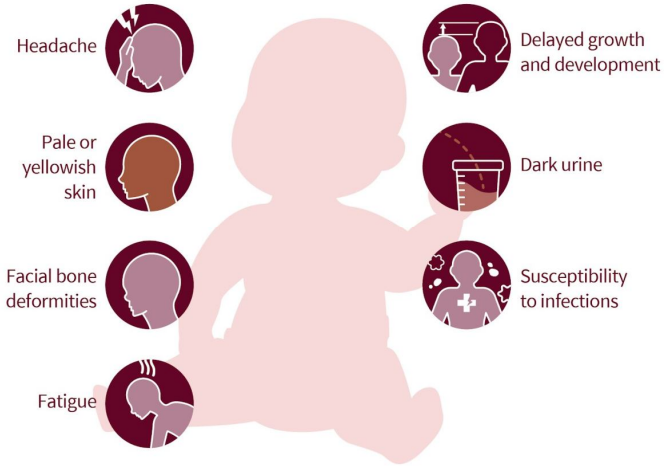
What is Thalassemia?



Malformed RBC in thalassemia



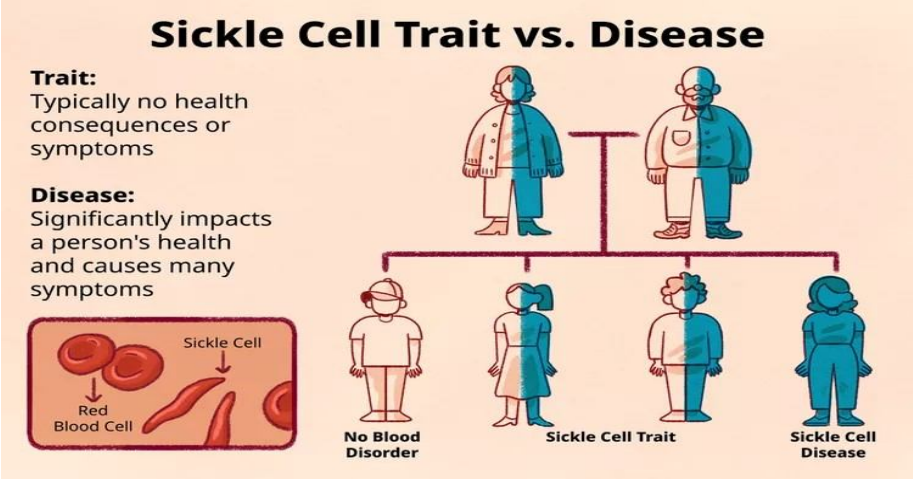
Clinical Features of Thalassemia



Role of Screening in Sickle Cell and Thalassemia

- The signs and symptoms of sickle cell anemia usually appear around 6 months
- But**,** People with **Sickle cell Trait** usually do not have **any of the symptoms** of sickle cell disease (SCD), but they can pass the trait on to their children.
- **Thalassemia disesase** are usually detected in **their childhood (usually diagnosed within the first two years of life)**
- People who have **thalassemia trait do not usually experience any health problems except perhaps a mild anemia** and live normal.
- It is important for individuals to be aware of their **thalassemia trait status**, **particularly individuals of reproductive age group.**
- So **identification of both disease & trait conditions** are important to address issues of Sickle cell and Thalassemia in the State**.**

Role of Screening in Sickle Cell and Thalassemia



Need of Elimination Programme

- The outcome of the existing projects in the State i.e "Odisha Sickle Cell Project" in 12 western districts and "Control of thalassemia and SCD in Odisha" in collaberation with CMC vellore is a matter of concern.
- Prevalance of Sickle cell is 15% in the state
- Prevalence of thalassemia is 5-10% in the state
- Social stigma and ignorance around the disease & trait conditions continue to remain a public health challenge in India including Odisha

Proposed strategies for elimination

- **GoI Target for elimination :Sickle Cell by 2047 & Thalassemia- Not defined**
- **Three pronged Strategies Proposed for elimination:**
  - **Universal screening** of all population (<= 25 years) for early identification of disease & carrier conditions.
  - **Implementation of Comprehensive prevention programs** which include public awareness and education, carrier counseling as well as information on prenatal diagnosis, post conception and pre-implantation diagnosis and genetic counseling etc.
  - Treatment & Management of cases with **Sickle Cell Anemia and Thalassemia disease**.

Target Population of screening

- Under National Sickle Cell Anaemia Elimination Programme, the universal screening of sickle cell target Population is 0-40 yrs. In the first phase 0-25 yr population will be screened in the 20 targeted districts. The screening target of Odisha in the FY 2023-24 is 23 lakhs approx.
- State has mandate to screen thalassemia along with Sickle Cell screening.

Strategy of screening

- Through Point of Care test kits for screening of Sickle Cell at field level i.e SC-HWCs, VHSND and AWC/School level.
- Opportunistic screening/screening of OPD referred cases through Gazelle at SDHs/CHC/PHCs and camp approach screening through MHT/MHU.
- Through HPLC at DHHs for Opportunistic screening/screening of OPD referred cases.
- Field level screening through DBS in Gajapati, Ganjam and Rayagada Districts.

Pre-Natal Counselling

- Motivating all ANC PW for screening
- Motivating spouse testing for Positive ANC PW.
- If both found positive, then the mother is advised for pre natal diagnosis.
- If the CV sampling is done at SCB MCH, VIMSAR Burla and SLNMCH, Koraput.
- If the foetus is found positive, then proper steps are advised after taking the proper consent of parents.

Pre-Marital Counselling

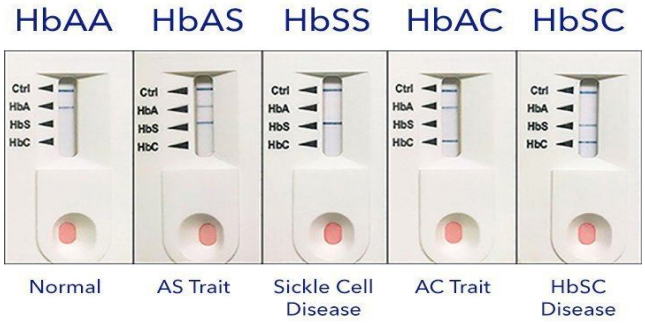
- Motivating all eligible couples for screening
- Encouraging Sickle trait person can marry normal ones as the sickle trait person with no symptoms can lead normal life.
- Advising that One SCD/carrier person shpuld not marry another SCD/carrier which may lead to SCD offsprings .

Importance of school/AWC screening

- Early diagnosis
- Early treatment for Positive patients
- Through regular follow up and medication the life expectancy can be increased for a child.
- Advising positive patient for BMT (Bone Marrow Transplantation)

Counselling for Opportunistic screening

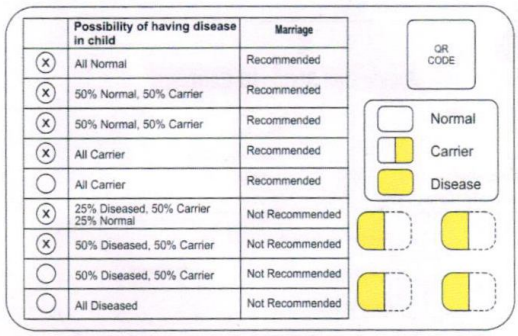
- The anaemic patients (Hb<9) must be encouraged for screening of SCD and thlassemia if not referred from OPD.



Sickle Cell Status card

- All screened persons will be availed the sickle cell card.
- 6 types of cards available
- Cards of Males are blue and those of females are pink in colour.
- The cards will be provided for AA(Normal), AS (carrier) and SS (SCD).

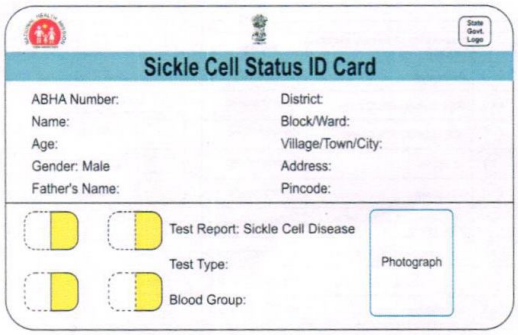
Sickle Cell Status card (Male)



	Possibility of having disease in child	Marriage
⊗	All Normal	Recommended
⊗	50% Normal, 50% Carrier	Recommended
⊗	50% Normal, 50% Carrier	Recommended
⊗	All Carrier	Recommended
○	All Carrier	Recommended
⊗	25% Diseased, 50% Carrier 25% Normal	Not Recommended
⊗	50% Diseased, 50% Carrier	Not Recommended
○	50% Diseased, 50% Carrier	Not Recommended
○	All Diseased	Not Recommended

QR CODE

Normal
Carrier
Disease



Sickle Cell Status ID Card

ABHA Number:
Name:
Age:
Gender: Male
Father's Name:

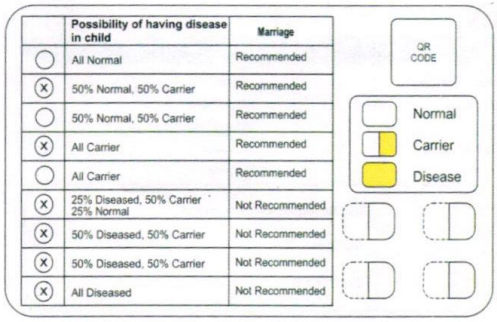
District:
Block/Ward:
Village/Town/City:
Address:
Pincode:

Test Report: Sickle Cell Disease
Test Type:
Blood Group:

Photograph

Sickle Cell Status card (Female)

Back



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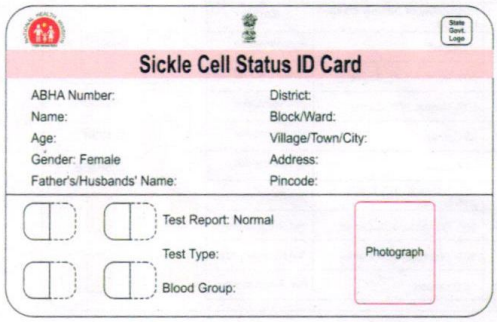
⊗	25% Diseased, 50% Carrier 25% Normal	Not Recommended
⊗	50% Diseased, 50% Carrier	Not Recommended
⊗	50% Diseased, 50% Carrier	Not Recommended
⊗	All Diseased	Not Recommended

QR CODE

Normal
Carrier
Disease

Front



Sickle Cell Status ID Card

ABHA Number:
Name:
Age:
Gender: Female
Father's/Husbands' Name:

District:
Block/Ward:
Village/Town/City:
Address:
Pincode:

Test Report: Normal
Test Type:
Blood Group:

Photograph

Matching of Sickle Cell Status card

- The society must be encouraged for matching of their sickle cell cards.
- The special cards are specially designed to predict the genotype pattern of their offspring after matching of the card.
- This may help the persons to take any suitable decision about their marriage.
- This will prevent the incidence of SCD in future.

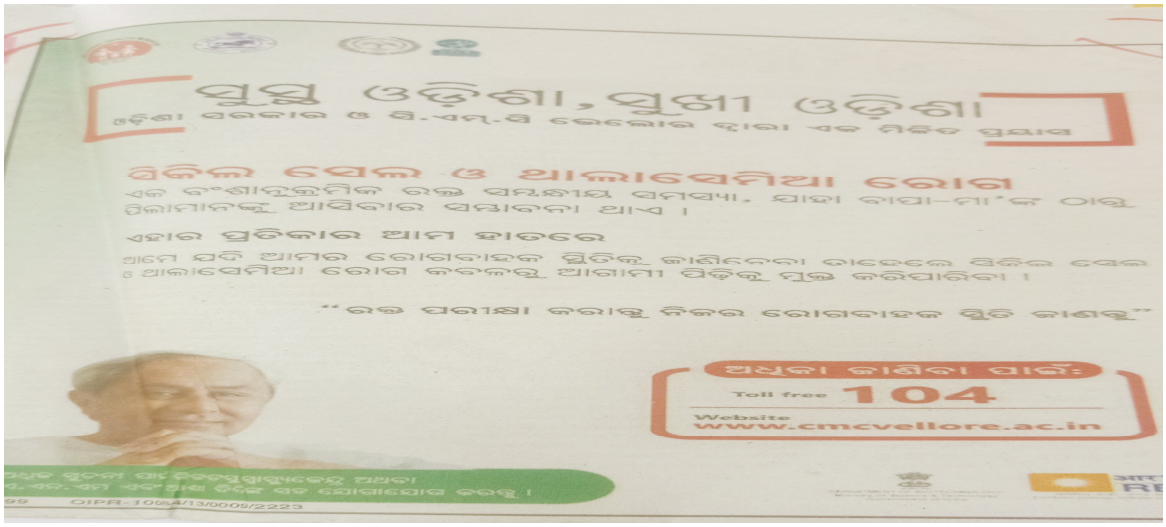
Thalassemia Bal Sewa Yojna (TBSY)

- CIL is one of the first PSUs to take CSR project for curative treatment of thalassemia.
- Financial assistance upto Rs.10 lacs is provided to ten prominent hospitals spread across the country for bone marrow transplants of eligible patients.
- Patient's age must be 12 years or less.
- Patient's family income should be Rs.8 lakhs per annum or less.
- Application to be submitted on TBSY Portal

Encouragement of other Govt schemes

- Rs.500 per month is provided to the patients of SCD and Thalassemia through DBT towards transportation cost after registration of the patient with all relavant documents at the nearest CHC.
- Free Blood (service charge exempted).
- Free drugs for SCD/thalassemia available at Govt. Hospitals.

Paper publication




"ସିକିଲ୍ ସେଲ୍ ଏବଂ ଥାଲାସେମିଆ ମୁକ୍ତ ଓଡ଼ିଶା"
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Thank You


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