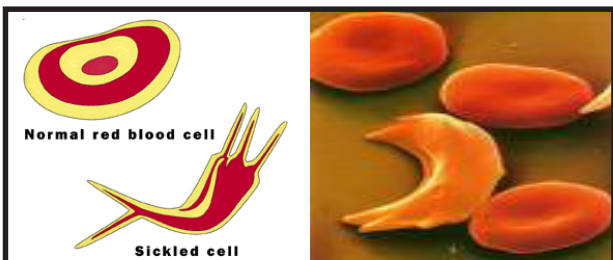


# SICKLE CELL DISEASE

With reference to Western Orissa

By : Dr. K.C.Dash M.D

Sickle Cell disease (SCD) is a “Blood disease” Human Blood which flows through out the body is made up of plasma and Cells. Out of the cells, Red Blood Cells (RBC) which transport Oxygen from the lungs to different tissues of the body through the Hemoglobin (Hb). This Hb are of different types, out of which Hb-A is found in adult mostly but when Hb ‘s’ predominates over the Hb-A then the sickle cell disease is manifested. The R.B.C. is usually round in shape



but due to presence of Hb ‘s’ it (RBC) becomes elongated and curved as a result of which it looks more or less like a “sickle” for which the disease is know as “Sickle cell disease”. It is a hereditary disease, neither “infective’ nor ‘Contagious”.

**Type :** The S.C.D. are of two types :**Sickle cell Disease** – Where Hb ‘s’ (Abdominal Hb) is predominant & almost majority of the RBC are sickle shaped permanently (irreversible)**Sickle cell Trail** – Here both normal Hb-A is predominantly present where as Hb ‘s’ (abdominal Hb) also present in 10-20% only. Here the condition is reversible & most of the RBC are

### About The Writer

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normal shape & a few sickled for which most of the patients are asymptomatic.

**Prevalance** - Sickle cell Disease was detected since long in Central Africa (20%), almost Negro population of America (8%) and India specially in Western Orissa but no authentic survey report is available in this respect. Many surveys by different N.G.O & voluntary organization were conducted in patchy manner. Hence a mass survey is required to know the exact position of the disease in the are.

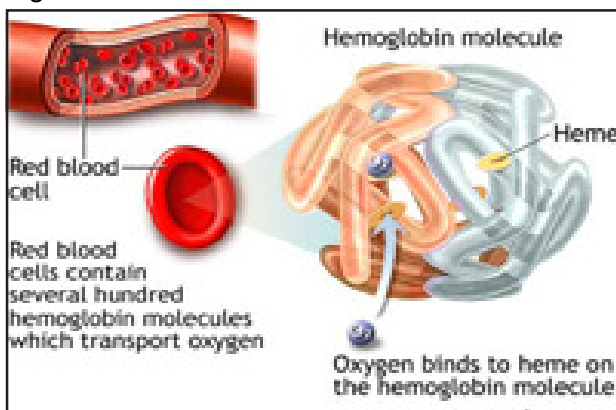
However from the patients attending to specially V.S.S. Medical College Hospital, Burla

**Sickle cell disease** is a genetic blood disorder which affects red blood cells. Normal red blood cells possess hemoglobin A, and this causes them to be soft and round. These physical characteristics allow them to fit through very small blood vessels without difficulty. Red blood cells containing hemoglobin A have a typical life span of about 120 days, after which they are replaced by new red

blood cells. Sickle cell disease causes production of a different type of hemoglobin, hemoglobin S. Hemoglobin S does not create the same soft, round shape that was seen with hemoglobin A. Instead the red blood cells have a sticky texture, and they may become stiff and sickle-shaped. These sticky or sickle-shaped blood cells are not able to pass as easily through the tiny blood vessels in the body, and they also have a short life span, typically less than 20 days.

and other Hospitals of the Western Orissa like Sundargarh, Sambalpur, Bargarh, Jharsuguda, Deogarh, Bolangir, Kalahandi and Phulbani are taken as 2 "SICKLE CELL BELT".

Among the tribal people some castes specially other Backward and Schedule Castes people the incidence of S.C.D. is predominantly high. But no caste is immune from this disease.



**Presenting symptoms :**

In SCD the presenting features are as follows which are found since childhood :Anaemia (Bloodlessness) mild jaundice. Retarded Growth.

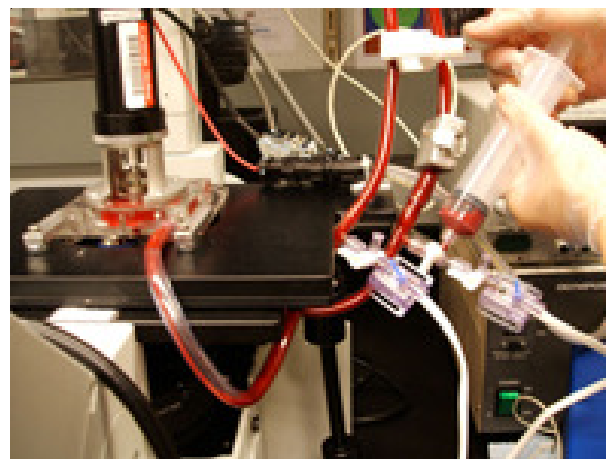
Delayed in the development of milestone in the childhood like delay in standing, walking, unable to run 7 sit property corresponding to their age. Protruded abdomen. Repeated ill health due to infection of the upper. Respiratory tract by various organisms as they are susceptible and vulnerable to different micro-organize. Breath – less ness during walking even on sitting specially in high attitude and while moving in air (Aero plane)Swelling of face and legs. Diffuse pain on

chest, abdomen, head, legs and different sites of the body.

**Diagnosis :**

SCD is not a single disease entity. It is a 'symptom complex' known as "SUNDROME" In presence of Haemolytic Anaemia. Intermittent episodes of pain at various sites of body. & above all –Demonstration of " Sickling Test" from a drop of Blood covered with cover ship (sealed) and detect sickle shaped RBC under ordinary microscope (with in 24-48 hrs)

However it is to be subsequently confirmed by "Hemoglobin electrophoresis"



specially for sickle cell trait for the presence of Hb –'s'

**Complications or crisis ;**

Patients of sickle cell disease (SCD) very often present with severe chest pain & admitted to cardiac unit (ICU), presenting as acute abdomen specially like Appendicitis, Renal colic or biliaycolic for which during the routing

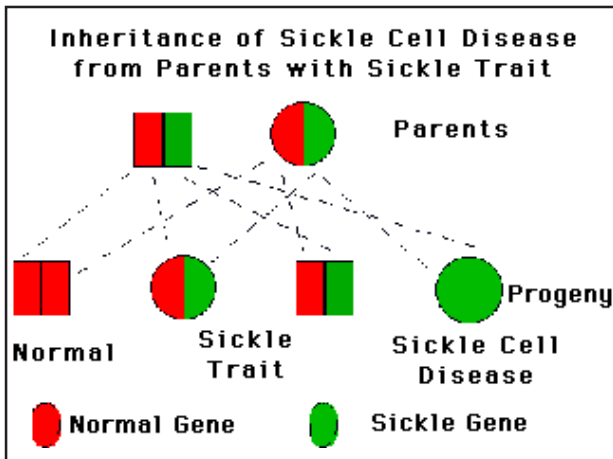
**Hemoglobin structure**

Hemoglobin is a conjugated metallo-protein with 4 globular polypeptide subunits; 2 alpha and 2 beta globins. Each globin contains a porphyrin ring structure called heme (a porphyrin ring is made of 4 Nitrogen containing pyrole groups held together by methyl bridges). Each heme group covalently binds a central ferrus iron (Fe<sup>2+</sup>). It is the ferrus iron (Fe<sup>2+</sup>) in the middle of each of the heme rings which can reversibly bind

O<sub>2</sub>. Of the 6 potential binding sites on each of the 4 Fe<sup>2+</sup>'s; 4 are bound to the porphyrin ring (the heme ring), 1 is bound to a histidine (the amino acid His) residue within the globin portion of the hemoglobin, and the remaining binding site is available to bind 1 molecule of O<sub>2</sub>, carbon monoxide (CO), or something else. Thus, each hemoglobin can carry 4 O<sub>2</sub>'s. One RBC contains 250 million Hb molecules (ie. 1 RBC can carry as many as 1 billion molecules of O<sub>2</sub>).

investigation. Blood sickling is to be examined as mandatory.

**Hemolytic Crisis** – Normal Red Blood Cell (RBC) survives for about 90 days but sickle RBC has short span of life for which due to rapid destruction of RBC in the spleen, blood becomes



haemolysed and manifested as jaundice & hemolytic Anaemias.

**Vasooclusion** – round shaped RBC flows freely inside the blood vessels & capillaries but the sickle RBC stick with each other as a result of which they were adherent and creates block age in different blood vessels and manifested as ischemic pain in different organs like bone, joints, even in eyes. Thereby causing fevers headache, vomiting etc.

**Sequestration Crisis :**

Due to venous obstruction of the spleen it is the first organ to face the crisis for which the children became the victim in their easy childhood due to repeated crisis. The kidney is also not

spared & faces renal necrosis leading to Renal failure in adult. The long bone like “Femur” & “Hemerus” are affected & aseptic necrosis and Osteomyelitis in the long bones are frequently seen. Likewise no organ is spared from the “sick Cell Crisis” as it is a Disease of the Blood.

**Treatment** There is no specific treatment for SCD. However folic acid, suitable Antibiotics are being used as a prophylactic measure. During the period of crisis the patient should be hospitalized to alleviate pain etc. and to combat Anaemia blood transfusion may be required. However to reduce the rate of S.C.D. the parents should be cautious while fixing marriages of their sons & daughters. Instead of going for the horoscope etc. they should select a bride or groom who should be free from “Sickle Cell” if their children are having the same since birth. By that their off springs will be partially spared from the disease marriage counseling in this respect is to be stressed in the society.

All the patients of SCD does not go to crisis specially the Traits. Who remain asymptomatic. Hence one should not be panic on this respect. In the winter one should protect from cold but not necessarily that he or she should drink less water. They may not take bath in cold water. Sponging with slight warm water is advised during the winter.

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**Navratna Info**

**Hemoglobin S (HbS).** HbS (S is for sickle) is the abnormal variant of hemoglobin A, which occurs in sickle-red blood cells and is the primary characteristic of the disease. The difference between hemoglobin A (HbA) and hemoglobin S (HbS) lies in only one protein out of about three hundred that are common to both. This protein lies along an amino-acid chain called beta-globin, where even a tiny abnormality has disastrous results.

· **Hemoglobin F (HbF).** HbF (F is for fetal) is a form of hemoglobin that is produced in everyone

during fetal development in the womb and for a short time after birth. Normally, most HbF is later replaced by hemoglobin A, although some HbF may persist throughout life. Importantly, HbF is able to block the sickling action of red blood cells. Infants who have inherited sickle cell disease, then, do not develop symptoms of the illness while they still have HbF. People with the sickle cell gene who continue to carry some fetal hemoglobin are better protected, therefore, from severe forms of the disease. It is being used as the basis for therapies used in sickle cell.