

PHARMA HEALTH CLUB

SAFE USE OF MEDICINES FOR BETTER HEALTH

1ST JULY, 2015

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WORLD SICKLE CELL DAY

June 19

'World Sickle Cell Day' is celebrated annually on 19th June. It was established by the United Nations General Assembly in 2008 in order to increase the awareness about the sickle cell disease and its cure among the common public. It was celebrated first time in 2009. Sickle cell disease has become a common and foremost genetic disease worldwide which is must to cure through awareness campaign, curable activities, early diagnosis and management. [1]

What is Sickle Cell Disease (SCD)?

- It is an inheritable, genetic and fatal disease causing red blood cells disorders which has been classified as sickle cell anemia and may lead to death. It is the most common public health problem in the African and Asian countries of the world. [3]
- SCD also known as Sickle-Cell Anemia (SCA) and drepanocytosis, characterized by an abnormality in the oxygen-carrying haemoglobin molecule in red blood cells. This leads to a propensity for the cells to assume an abnormal, rigid, sickle-like shape under certain circumstances. [5]

Sickle Cell Disease (SCD) is found in which areas?

Sickle cell disease (SCD) affects millions of people throughout the world and is particularly common among those whose ancestors came from sub-Saharan Africa; Spanish-speaking regions in the Western Hemisphere (South America, the Caribbean, and Central America); Saudi Arabia; India; and Mediterranean countries such as Turkey, Greece, and Italy. [6]

How Sickle Cell Anaemia develops in Human body ?

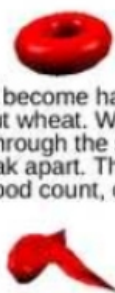
- Normal red blood cells are quite elastic, which allows the cells to deform to pass through capillaries. [5]
- The loss of RBC elasticity is central to the pathophysiology of sickle-cell disease. In sickle-cell disease, low-oxygen tension promotes RBC 'Sickling' (Cell narrows and becomes C-shaped) and repeated episodes of sickling damage the cell membrane and decreases the cell's elasticity. [5]
- These cells fail to return to normal shape when normal oxygen tension is restored.

- As a consequence, these rigid blood cells are unable to deform as they pass through narrow capillaries, leading to vessel occlusion and ischemia. [5]
- The actual anemia of the illness is caused by haemolysis, the destruction of the red cells, because of their shape. Although the bone marrow attempts to compensate by creating new red cells, it does not match the rate of destruction. Healthy red blood cells typically function for 90-120 days, but sickled cells only last 10-20 days. [5]

Sickle Cell Anemia

Sickle Cell anemia is an inherited red blood cell disorder. Normal red blood cells are round like doughnuts, and they move through small blood tubes in the body to deliver oxygen.

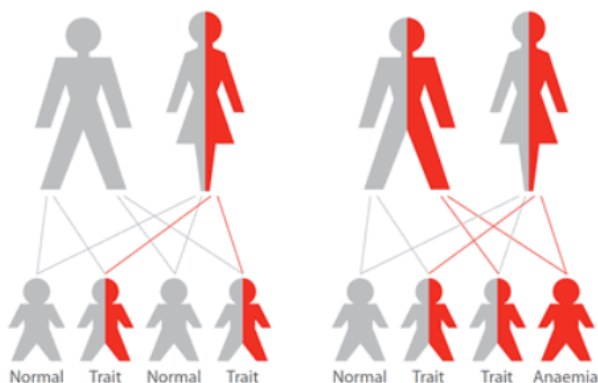
Sickle red blood cells become hard, sticky and shaped like sickles used to cut wheat. When these hard and pointed red cells go through the small blood tube, they clog the flow and break apart. This can cause pain, damage and a low blood count, or anemia.



TRANSMISSION

- Sickle cell disease is an INHERITED condition. [8]
- Two genes for the sickle hemoglobin must be inherited from one's parents in order to have the disease. [8]
- A person who receives a gene for sickle cell disease from one parent and a normal gene from the other has a condition called "Sickle Cell Trait." [8]

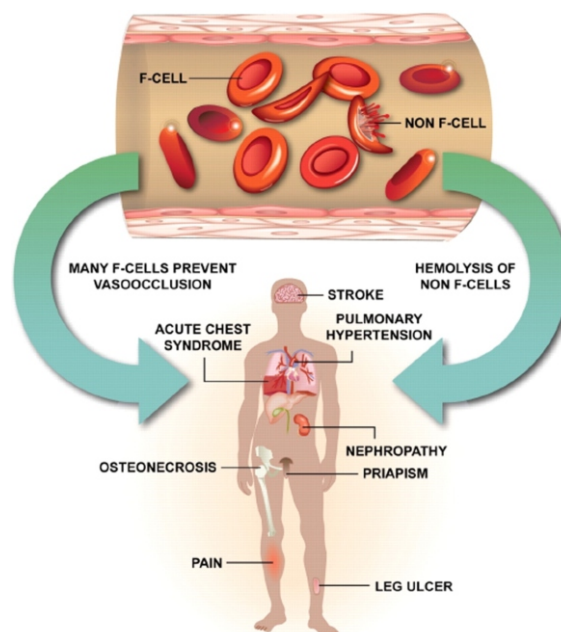
How the trait is passed on



SYMPTOMS [10]

- Severe pain
- Anemia
- Chest pain and difficulty breathing
- Strokes
- Joint pain and arthritis and bone infarctions
- Blockage of blood flow in the spleen or liver
- Splenomegaly
- Severe infections

DIAGNOSIS [10]



- Hemoglobin electrophoresis:** By measuring the amount of the abnormal sickle hemoglobin.
- Prenatal testing for SCD:** By examining the DNA of fetal cells obtained by chorionic villus sampling or amniocentesis.
- Complete Blood Count (CBC):** It reveals haemoglobin levels in the range of 6-8 g/dl with a high reticulocyte count (as the bone marrow compensates for the destruction of sickled cells by producing more red blood cells). Sickling of the red blood cells, on a blood film. [5]

Phenotype	Genotype	Positions to which hemoglobins have migrated	Origin	Hemoglobin types present
Sickle-cell trait	Hb^S / Hb^A			S and A
Sickle-cell anemia	Hb^S / Hb^S			S
Normal	Hb^A / Hb^A			A

Migration

HOW TO MANAGE SCD?

- **Folic acid and penicillin:** A 1 mg dose of folic acid daily for life. From birth to five years of age, they also have to take penicillin daily due to the immature immune system that makes them more prone to early childhood illnesses. [5]
- **Opioid pain medications:** (Ex.morphine) & NSAIDs (Ex.: ibuprofen) for management of vaso-occlusive crises (The vaso-occlusive crisis is caused by sickle-shaped red blood cells that obstruct capillaries and restrict blood flow to an organ resulting in ischemia, pain, necrosis, and often organ damage.). [5, 10]
- **Oxygen supplementation:** for hypoxia and in condition of Acute chest crisis (Acute chest syndrome (ACS) is defined by at least two of the following signs or
- **symptoms:** chest pain, fever, pulmonary infiltrate or focal abnormality, respiratory symptoms, or hypoxemia.) [5, 10]
- **Hydroxyurea:** FDA-approved medication that prevents painful episodes in sickle cell disease. Transfusion therapy: Blood transfusions are often used in the management of sickle-cell disease in acute cases and to prevent complications by decreasing the number of red blood cells (RBC) that can sickle by adding normal red blood cells. [5, 10]
- **Bone marrow transplants or Stem cell transplant:** It has been proven effective in children. Bone marrow transplants are the only known cure for SCD. [5] Stem cell transplant has a 5%-10% risk of death, but patients with successful transplants were completely cured of sickle cell disease, with no further episodes of pain. [10]?

AYURVED & SCD

Ayurved manages SCD in 3 steps[16]:

- 1.Dhatu Pushti Chikitsa - Treatment to strengthen body tissues and assist formation of hemoglobin through herbal medicines
2. Sroto Shodhan Chikitsa - Treatment to cleanse body channels of ama (toxins) so that the nutrient plasma could flow properly
3. Rasayana Chikitsa - Rejuvenative treatment aimed at improving energy levels and maintaining overall health

Medicinal treatment[17]:

Amla, Jethimadh, Majith, Haldi, Bavchi, Lodhra, Saatodi, Bhaangro and Galo are the main herbs used in SCD. Dadimadi Ghrita, Draksha Ghrita, Gomutra Haritaki and Shilajit are some of the important formulations for SCD.

Yograj Guggul and Laakshadi Guggul are two popular, clinically-approved Ayurvedic formulations for SCD.[18]

Diet[17]:

Diet containing folic acid and thiocyanate - Cabbage (Gobi), Cauliflower (Gobi ful) and Raddish (Mula), should be increased in the diet.

Nicosan, a phytochemical useful in sickle cell anemia is present in Sorghum (Jwar) and Clove (Lavang). Nitriloxide, another phytochemical, is present Sweet Potato (Sakariya), Spinach (Palak) & Moong (Mug), which can help in SCD.

Eight glasses of water everyday will help prevent dehydration.

Precautions[17]:

Avoid nasal decongestants. Avoid extreme heat or cold. Do not climb high altitudes. Reduce stress. Regular hematological and medical checkups are compulsory.

AYURVED MANAGES SCD IN 3 STEPS



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