babycell™

The Life Saving Cord Blood Bank


SICKLE CELL DISEASE

What is Sickle Cell Disease (SCD)?

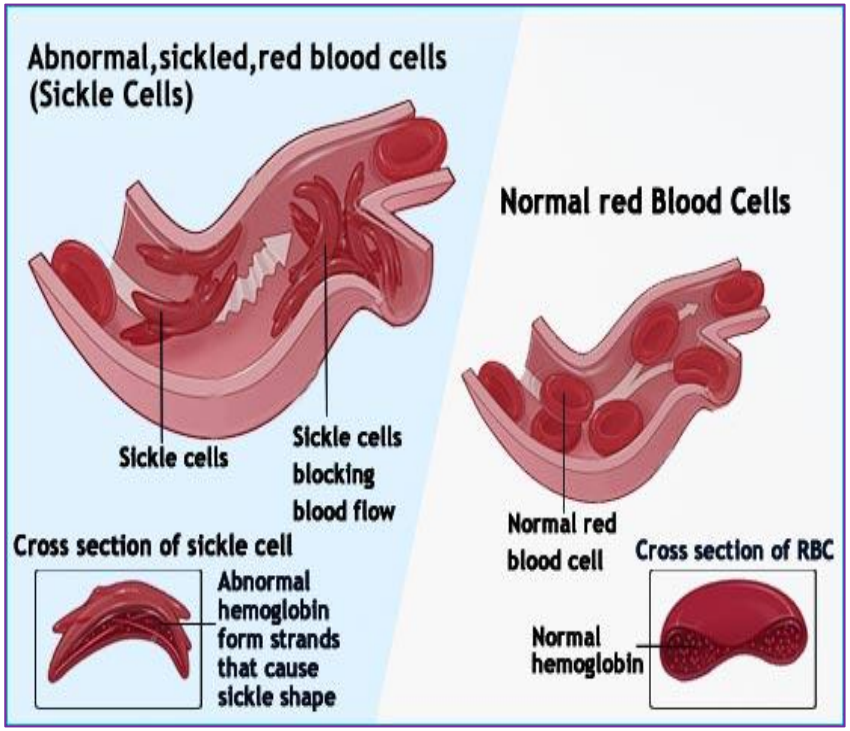
- *Sickle Cell Disease* (SCD) is a disease passed down through families in which red blood cells (oxygen carrying cells) form an abnormal crescent shape

- Children with SCD suffer from severe infections and damage to the organs in the body

- Some are even frequently hospitalized

What is Sickle Cell Disease (SCD)?

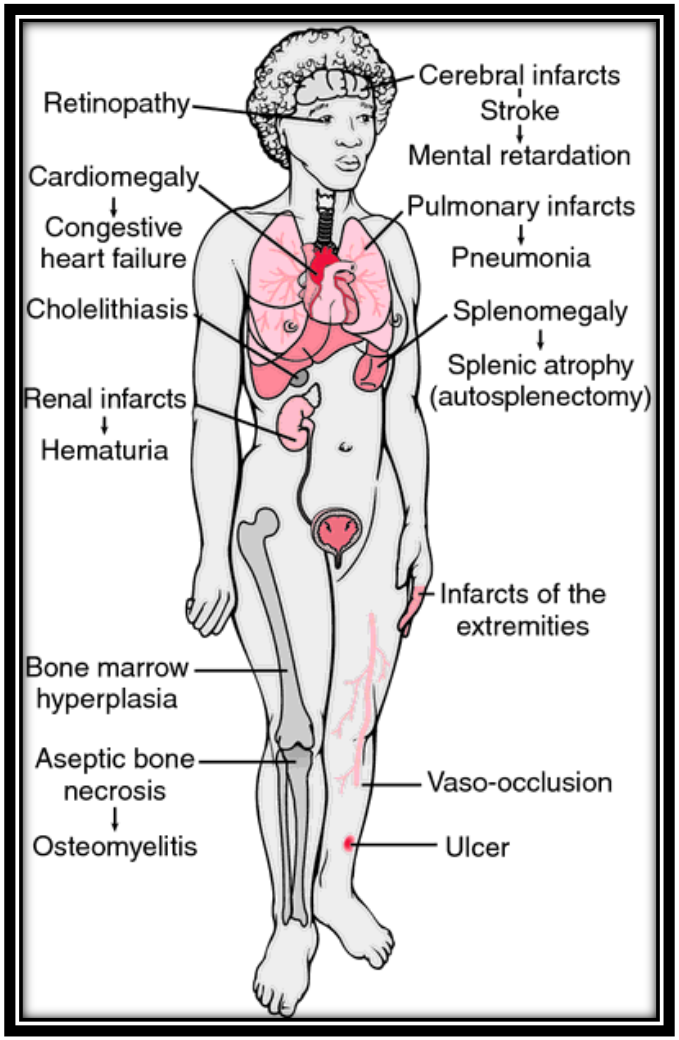
- It is an inherited blood disorder characterized primarily by chronic anaemia and periodic episodes of pain
- Haemoglobin molecules, in each red blood cell, carry oxygen from the lungs to body organs and tissues and bring carbon dioxide back to the lungs
- In sickle cell disease, ***the haemoglobin is defective***
- After haemoglobin molecules give up their oxygen, some may cluster together and form long, rod-like structures. These structures cause ***red blood cells to become stiff and sickle shaped***



Types of SCD

- There are several types of SCD
- The most common type is **Sickle Cell Anaemia** where a child has inherited two genes that produce an abnormal haemoglobin called "S" haemoglobin ("SS" disease)
- There are several other, less common types of sickle cell disease where one gene produces "S" haemoglobin and the other gene produces "C" haemoglobin **(SC disease)**
- **S-Beta Thalassemia** is caused when a child inherits one gene producing a "beta-thalassemia" type of haemoglobin and the other gene produces "S" hemoglobin ("S-beta thalassemia")
- Your child could have inherited only one of these types. It is important for you to know which one

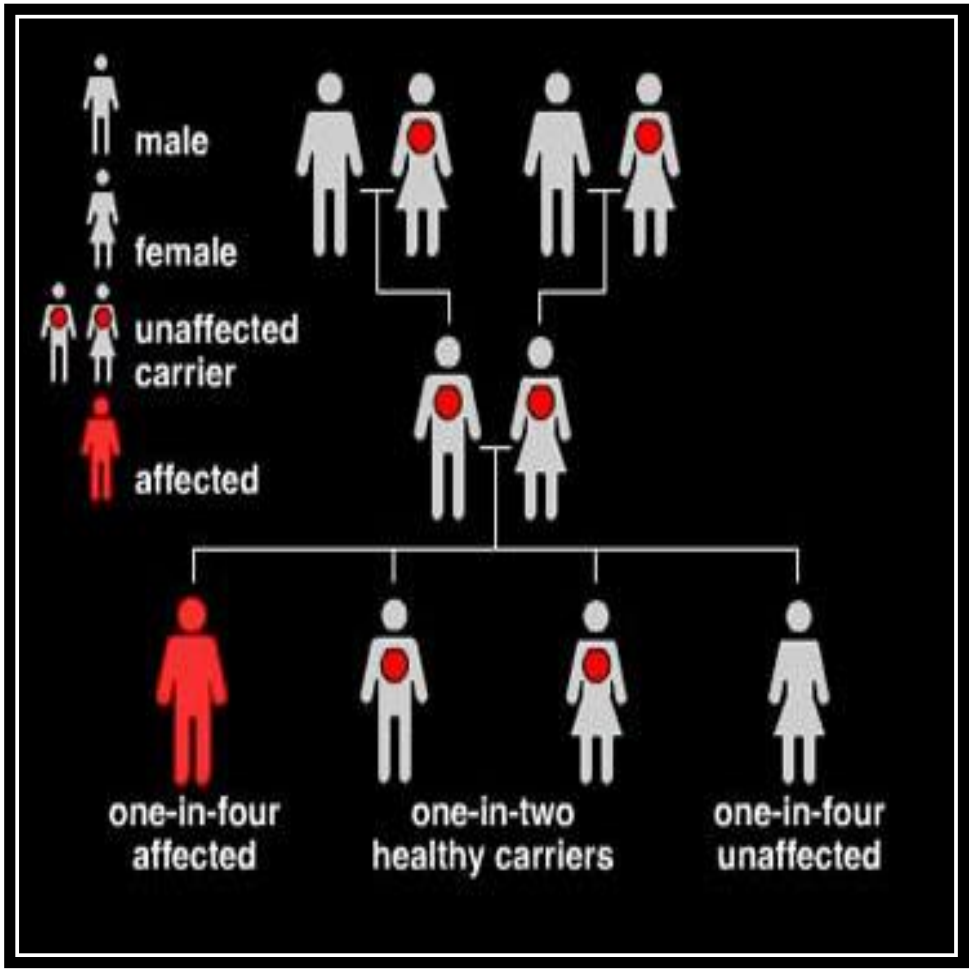
Symptoms of SCD



- Attacks of abdominal pain
- Bone pain
- Breathlessness
- Delayed growth and puberty
- Fatigue
- Fever
- Paleness
- Rapid heart rate
- Ulcers on the lower legs (in adolescents and adults)
- Yellowing of the eyes and skin (jaundice)

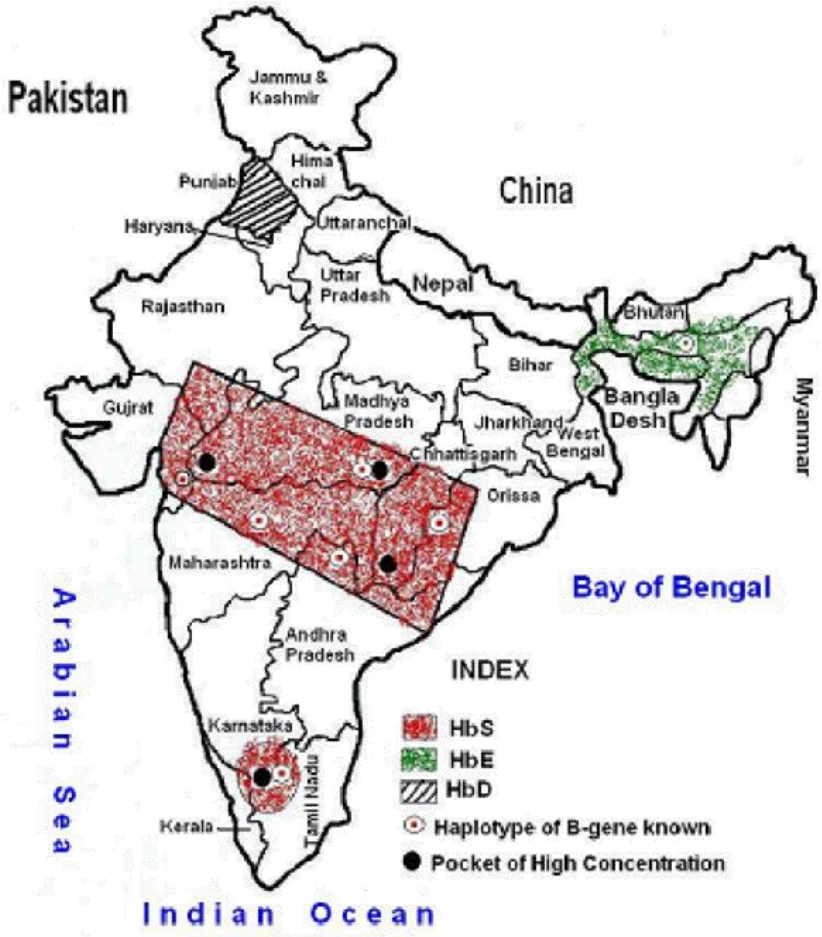
How does a child get SCD?

- The presence of two defective genes is needed for sickle cell disease



- If each parent carries one sickle haemoglobin gene (S) and one normal gene (A), each child has
  - a 25% chance of inheriting two defective genes and having sickle cell anaemia;
  - a 25% chance of inheriting two normal genes and not having the disease;
  - and a 50% chance of being an unaffected carrier like the parents

Incidence of SCD in India



- Highest incidence in Central and South India
- 10% of Indian population affected
- 40% are CARRIERS
- Main ethnicities affected include Punjabis, Parsis, Biharis and tribal populations across the country

Cord Blood Treatment

Successful Cord Blood Transplantation for Sickle Cell Anemia From a Sibling Who Is Human Leukocyte Antigen-Identical: Implications for Comprehensive Care

Gore, Lia M.D.; Lane, Peter A. M.D.; Quinones, Ralph R. M.D.; Giller, Roger H. M.D.; Journal of Pediatric Hematology/Oncology: September/October 2000 - Volume 22 - Issue 5 - pp 437-440

We report the successful transplantation of umbilical cord blood stem cells from a sibling who is human leukocyte antigen-matched to a child with sickle cell anemia. Conditioning was with busulfan, cyclophosphamide, and antithymocyte globulin. Time to neutrophil count >500/µL was 23 days and to platelet count >50,000/µL was 49 days. Full donor engraftment was achieved without graft-versus-host disease. This case demonstrates the potential usefulness of harvesting cord blood from full siblings of patients with sickle cell disease. Routine collection of **umbilical cord blood from siblings should be considered for patients with sickle cell disease**, and may increase acceptance and use of transplantation by families.

Cord Blood Treatment

Matched-related donor transplantation for sickle cell disease: report from the Center for International Blood and Transplant Research

Julie A. Panepinto et al; British Journal of Haematology Volume 137, Issue 5, pages 479–485, June 2007

Summary

We report outcomes after myeloablative haematopoietic cell transplantation (HCT) from human leucocyte antigen (HLA)-matched sibling donors in 67 patients with sickle cell disease transplanted between 1989 and 2002. The median age at transplantation was 10 years and 67% of patients had received >10 red blood cell transfusions before HCT. Most patients achieved haematopoietic recovery and no deaths occurred during the early post-transplant period. Sixty-four of 67 patients are alive with 5-year probabilities of disease-free and overall survival of 85% and 97% respectively. **This report confirms and extends earlier reports that HCT from HLA-matched related donors offers a very high survival rate, with few transplant-related complications** and the elimination of sickle-related complications in the majority of patients who undergo this therapy

Advantages of Cord Blood Banking

- Unless your baby has a sibling, SCD can only be treated via BONE MARROW TRANSPLANT
- This transplant requires a **100% matched donor**
- Probability of finding a perfectly matched donor = **1 in 30,000**

Advantages of Cord Blood Banking

- Banking cord blood can provide a source of treatment of SCD for your baby
- Cord Blood Banking is a painless, simple procedure
- It provides 80% chance of potentially curing your baby of SCD and 80 other blood-related disorders

THANK YOU

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