# OSCAR SANDWELL CO LTD

Organisation for Sickle Cell Anaemia Research and Thalassaemia Support

# Overview of Sickle Cell Anaemia & Thalassaemia Disorders

# What is Blood?

Sickle cell and Thalassaemia are blood disorders so it important to know the function of the blood in order to understand the conditions.

Blood is part of our body. It is pumped around our body by the heart.

It is made up of a liquid called Plasma and three types of cells.

The Plasma carries water, salts and material such as food to your tissues and then transports out waste products.

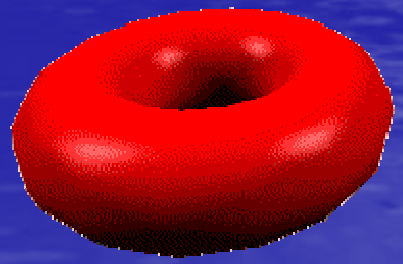
Your white blood cells defends your body against infections.

Your platelets stop you loosing blood if you hurt yourself, because they stick together blocking the cut to stop bleeding.

Then you have the red blood cells. The red blood cells carry oxygen around your body.

# What is the function of the Red Blood Cell?

- Carry oxygen from the lungs to the tissues as we inhale
- Carry carbon dioxide back to the lungs and out as we exhale
- Normally live 120 days
- Contains the protein haemoglobin, which is red and that is what makes our blood red
- Made from iron, folic acid, vitamin B12
- Made in the bone marrow



# Diagnosis

- Diagnosis is made by obtaining a sample of blood which is tested in a specific way to find out whether the person concerned has sickle cell trait or sickle cell disease and Thalassaemia trait or Thalassaemia disease. The test is called Heamoglobinopathy Screen.

# WHAT IS SICKLE CELL ANAEMIA?

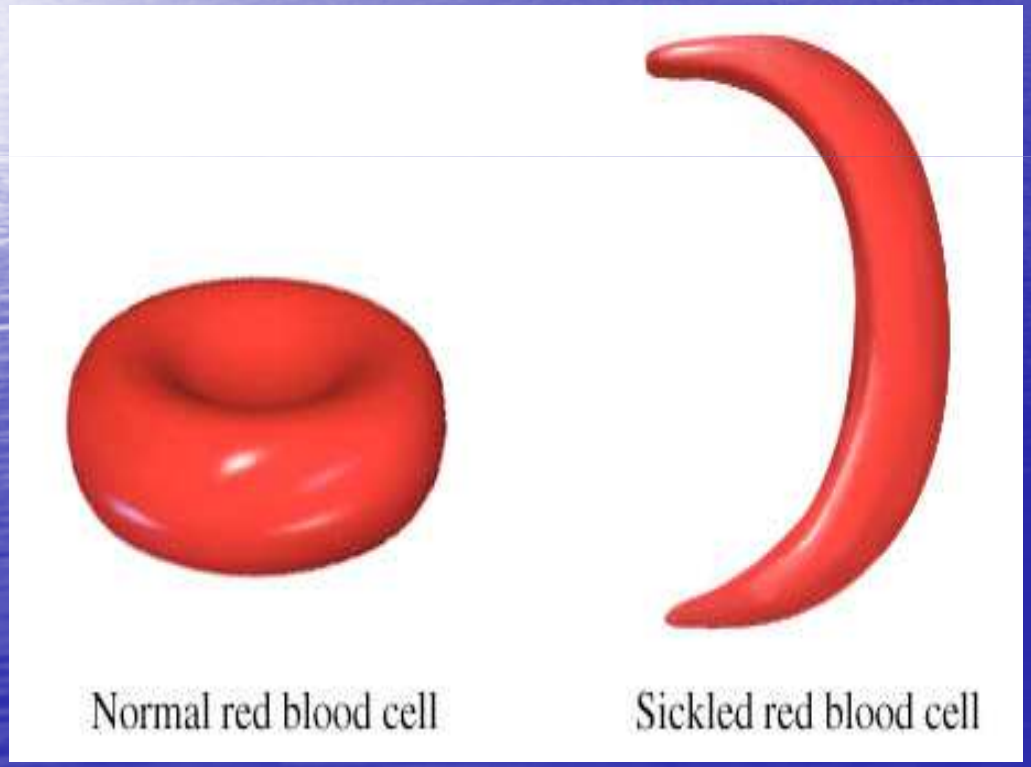
Sickle Cell Anaemia is an inherited blood

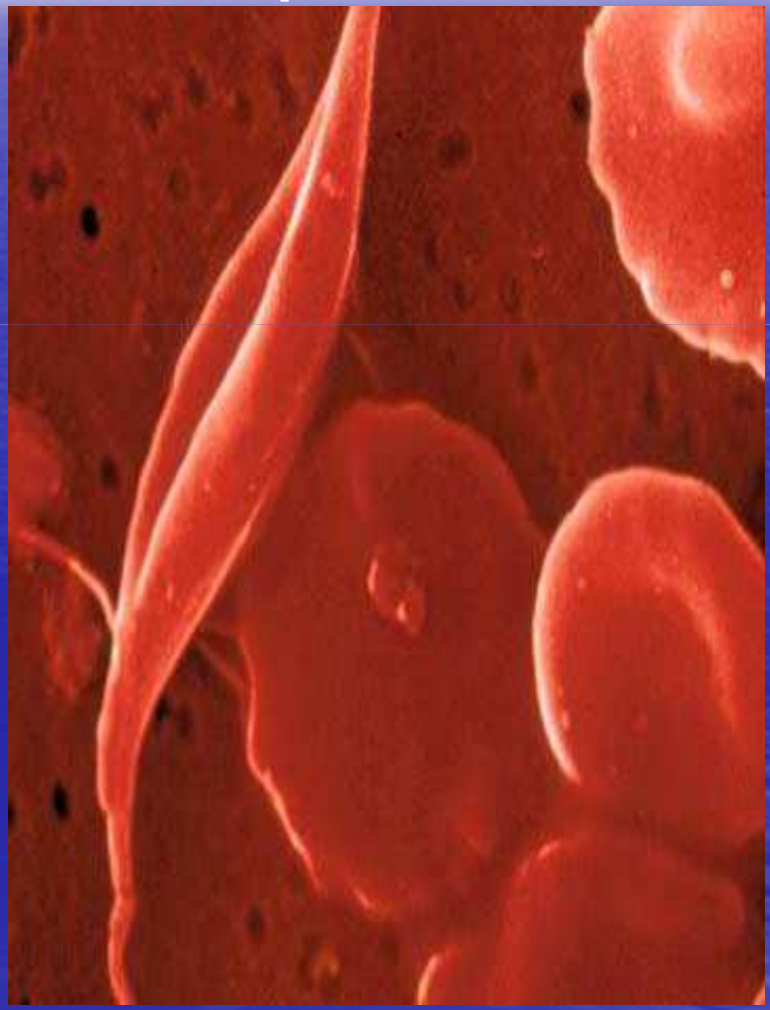
disorder, which affects the red blood cell.

It affects millions of people worldwide. Sickle cell anemia is most common among people whose ancestors come from Africa; Mediterranean countries such as Greece, Turkey, and Italy; the Arabian Peninsula; India; and Spanish-speaking regions in South America, Central America, and parts of the Caribbean. But it is not exclusive to these ethnicities.

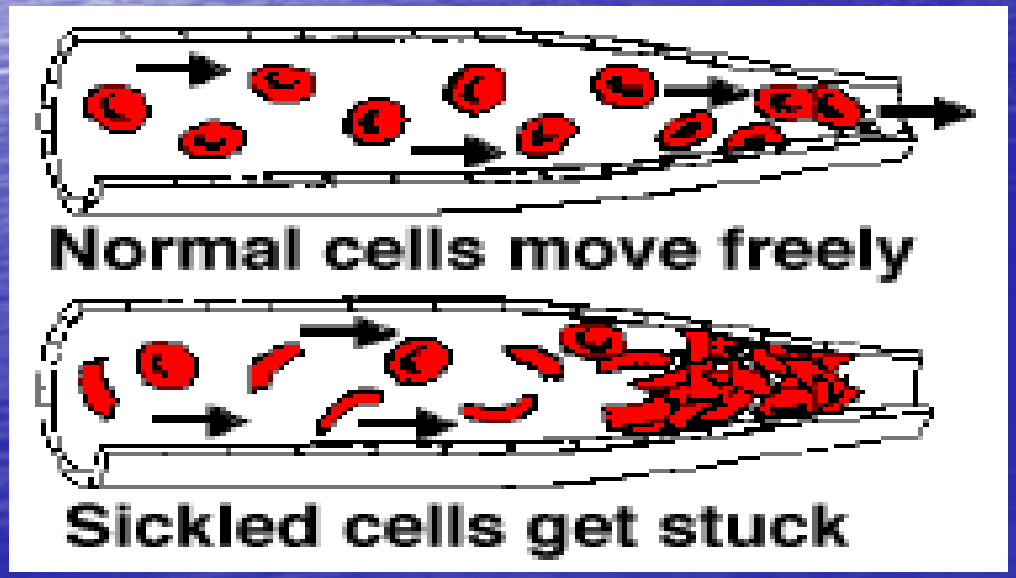
It is important to note Sickle Cell Anaemia is NOT contagious.

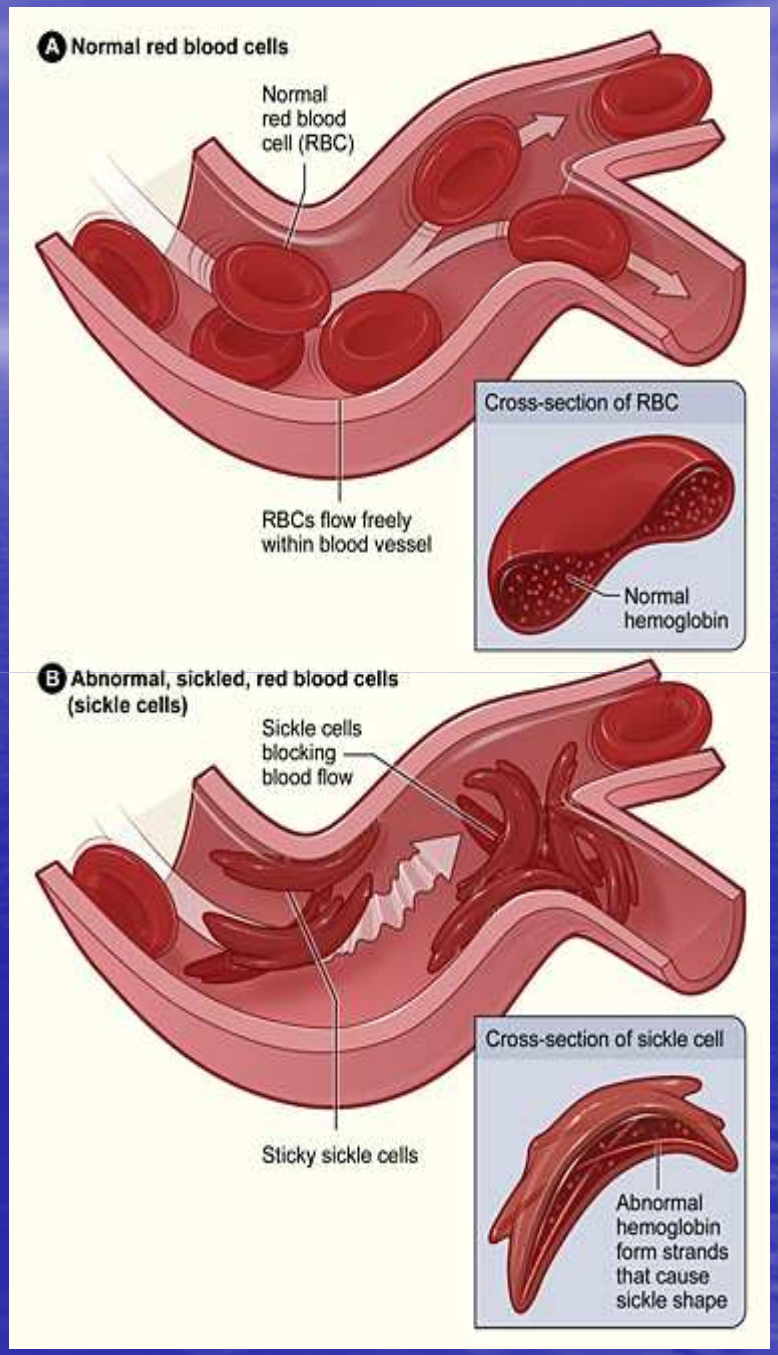
The reason the condition is called Sickle Cell is because the blood cells that carry oxygen around the body is in a sickle shape.



Normal haemoglobin cells are smooth and round, allowing movement through blood vessels. Sickle cell haemoglobin tend to cluster together, and cannot easily move through the blood vessels. This causes a blockage and stops the movement of oxygen-carrying blood. If there is no oxygen, then pain and damage occurs.



# SICKLE CELL TRAIT

- Sickle cell trait is the carriers state. The individual will carry one normal haemoglobin (**HbA**) and one abnormal e.g. (**HbS) = HbAS** (**Trait).**
- People with Sickle Cell trait lead completely normal lives and should not suffer any problems under normal circumstances.
- Sickle Cell trait cannot turn into **Sickle Cell Anaemia**, but can be passed on from parent to child.
- **Estimated 310,000 carriers in the UK**

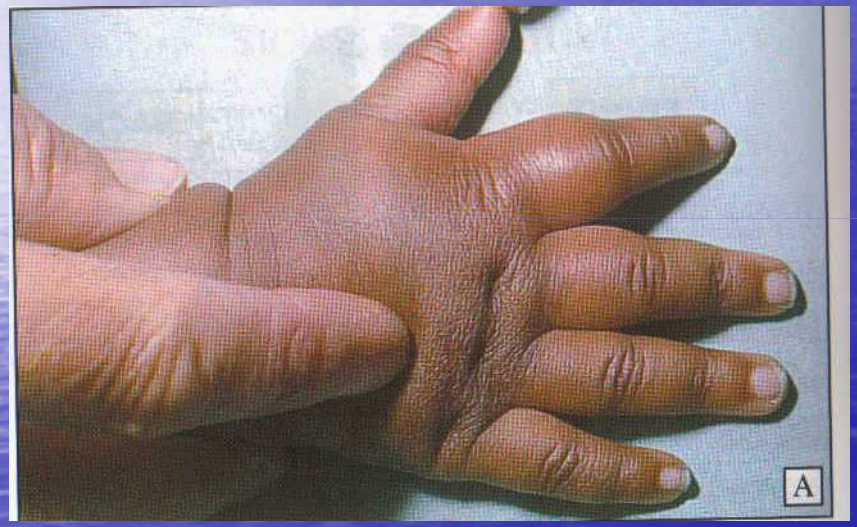
# Painful crisis

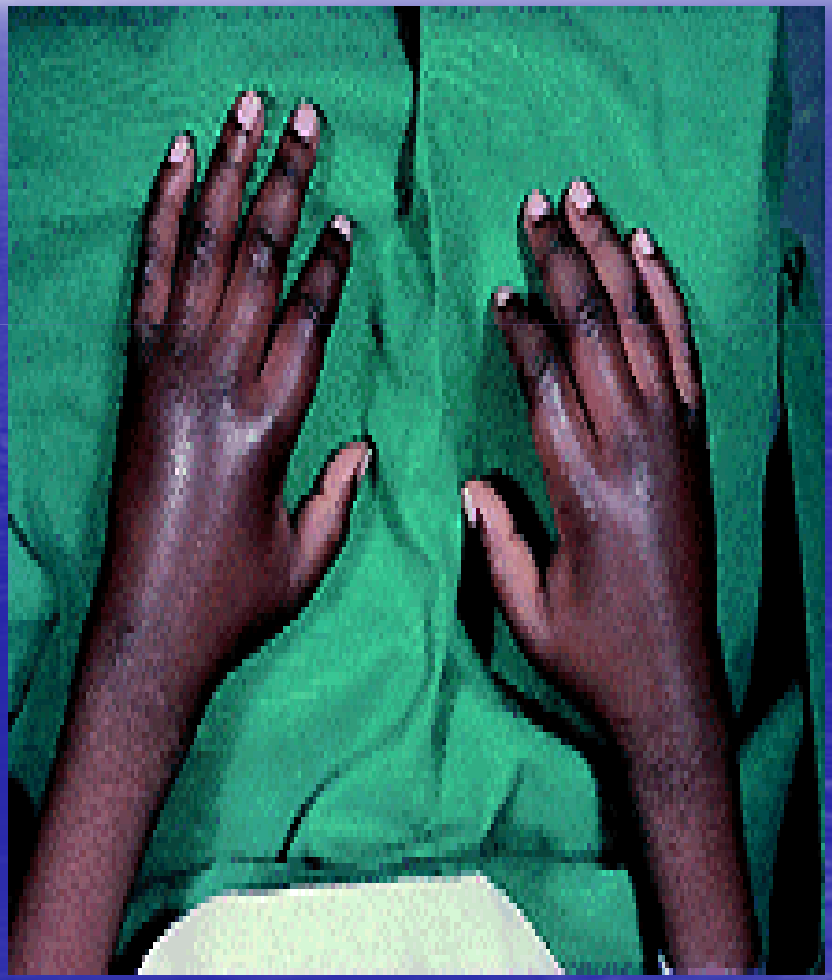
- The pain can occur anywhere, but most often occurs in the chest, arms, and legs. Painful swelling of the fingers and toes, called dactylitis, can occur in infants and children under 3 years of age.
- Priapism is a painful sickling that occurs in the penis. Any interruption in blood flow to the body can result in pain, swelling, and possible death of the surrounding tissue not receiving adequate blood and oxygen.

# Complications of Sickle Cell

- Anaemia
- Jaundice
- Painful/sickle crisis
- Strokes
- Increased infections
- Leg ulcers
- Bone damage
- Early gallstones
- kidney failure
- Eye problems
- Enlarge spleen

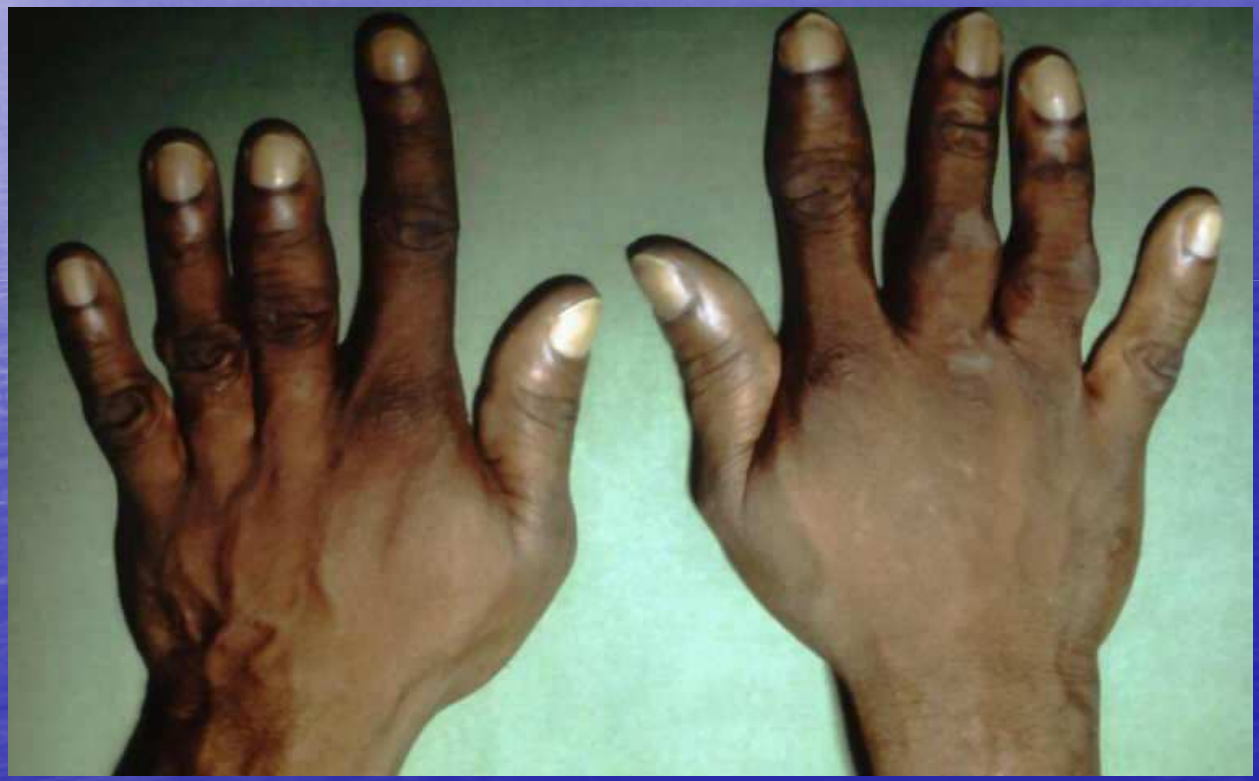
- Any and all major organs are affected by sickle cell disease. The liver, heart, kidneys, eyes, bones, and joints can suffer damage.



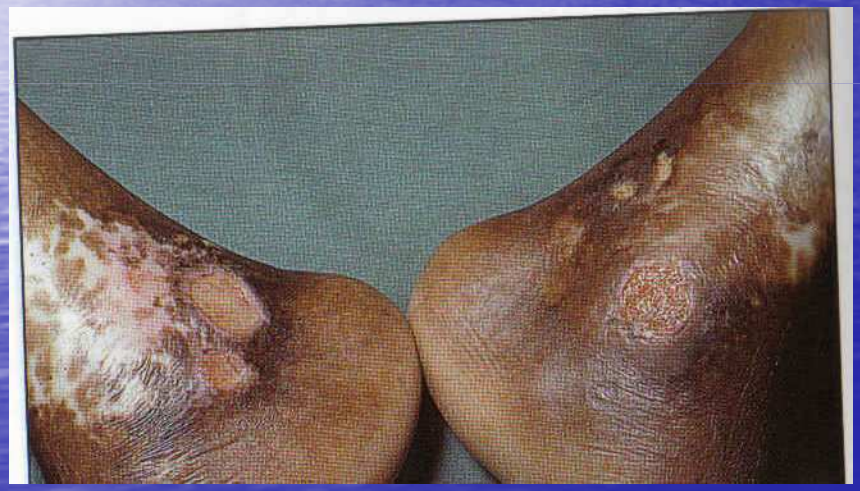



**DACTYLITIS**

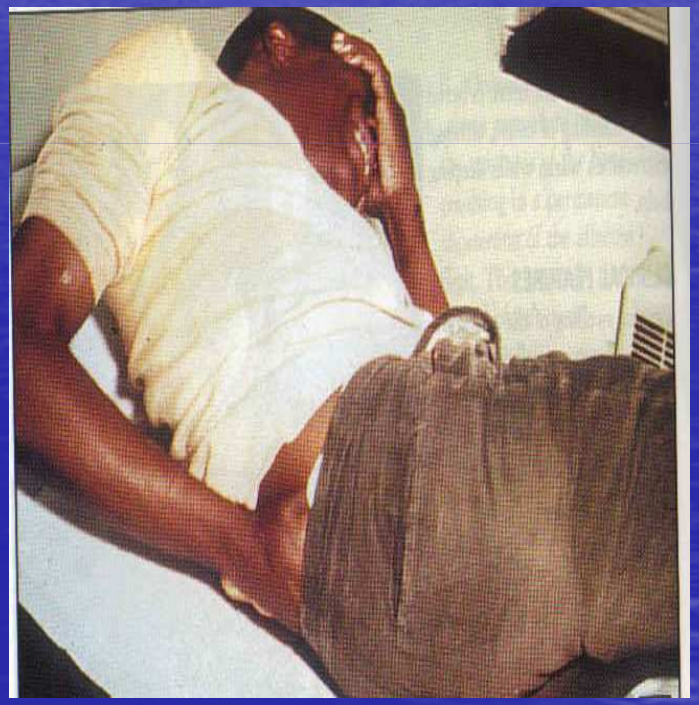
# Effects of dactylitis



## LEG ULCERS



## CRISIS IN SPINE



**DEEP JAUNDICE**

**AVASULAR NECROSIS OF FLAT BONES**


*(photo courtesy Dr. Klaas Wierenga, Kingston, Jamaica)*

# Managing SCD

- **Pain medication** for Sickle Cell Crisis.
- **Fluids** drinking plenty of water to prevent and treat crisis.
- **Rest**
- **Antibiotics** Penicillin to prevent infections
- Avoid hot/cold tempretures
- Healthy life style
- Avoid stress and overexertion
- Folic acid to prevent severe anaemia.

TREATMENT

- Blood transfusion to correct anaemia and prevent reoccurring strokes in children at high risk.
- Hydroxyurea
- Folic Acid
- Bone Marrow Transplant

# Any Questions

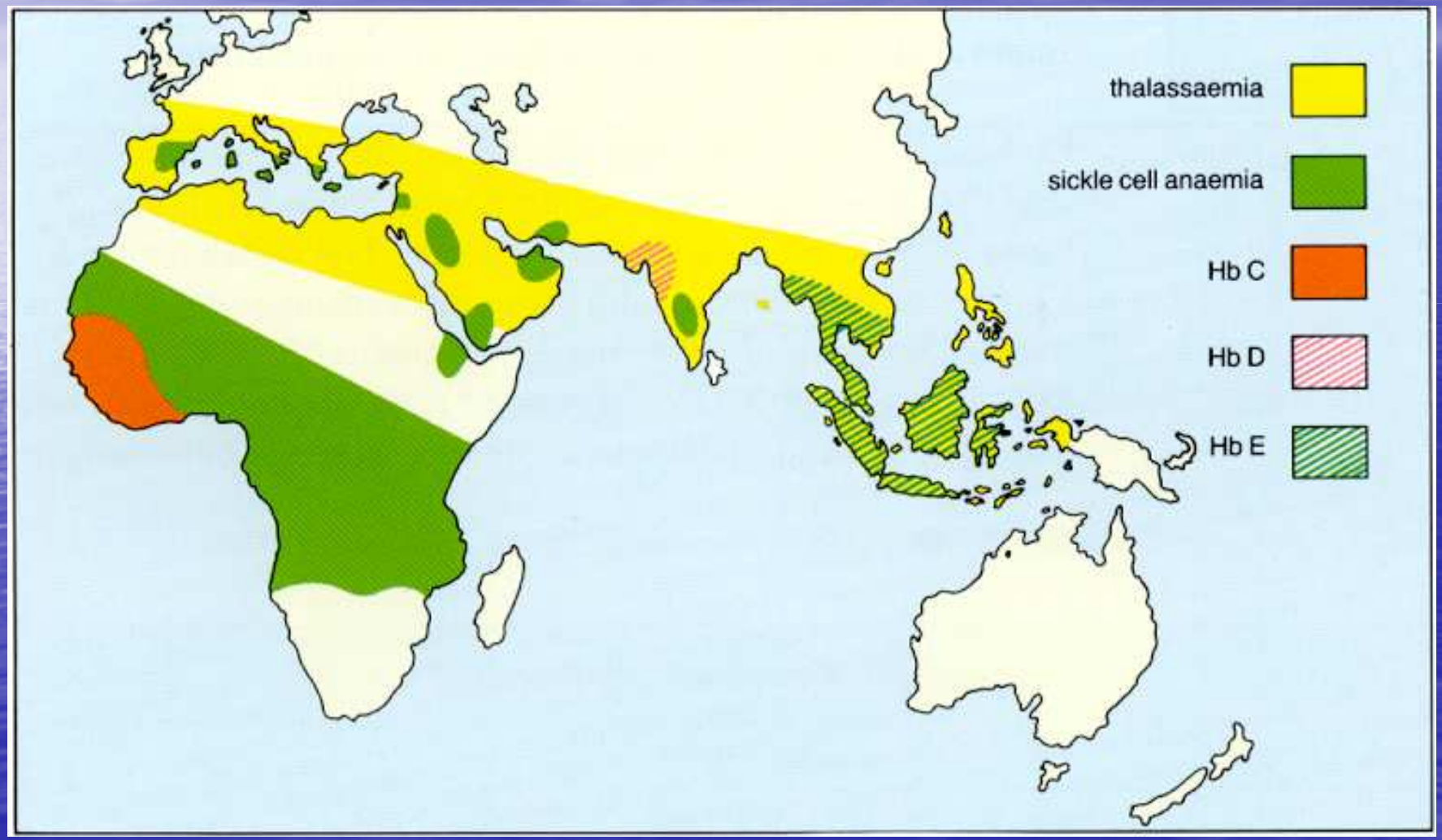
# WHAT IS THALASSAEMIA?

- Thalassaemia is a blood disorder that is found in many countries around the world, and particularly in people of Mediterranean, Middle Eastern or Asian origin. But as people have moved out of those regions we find there are many people in the UK living with the conditions.

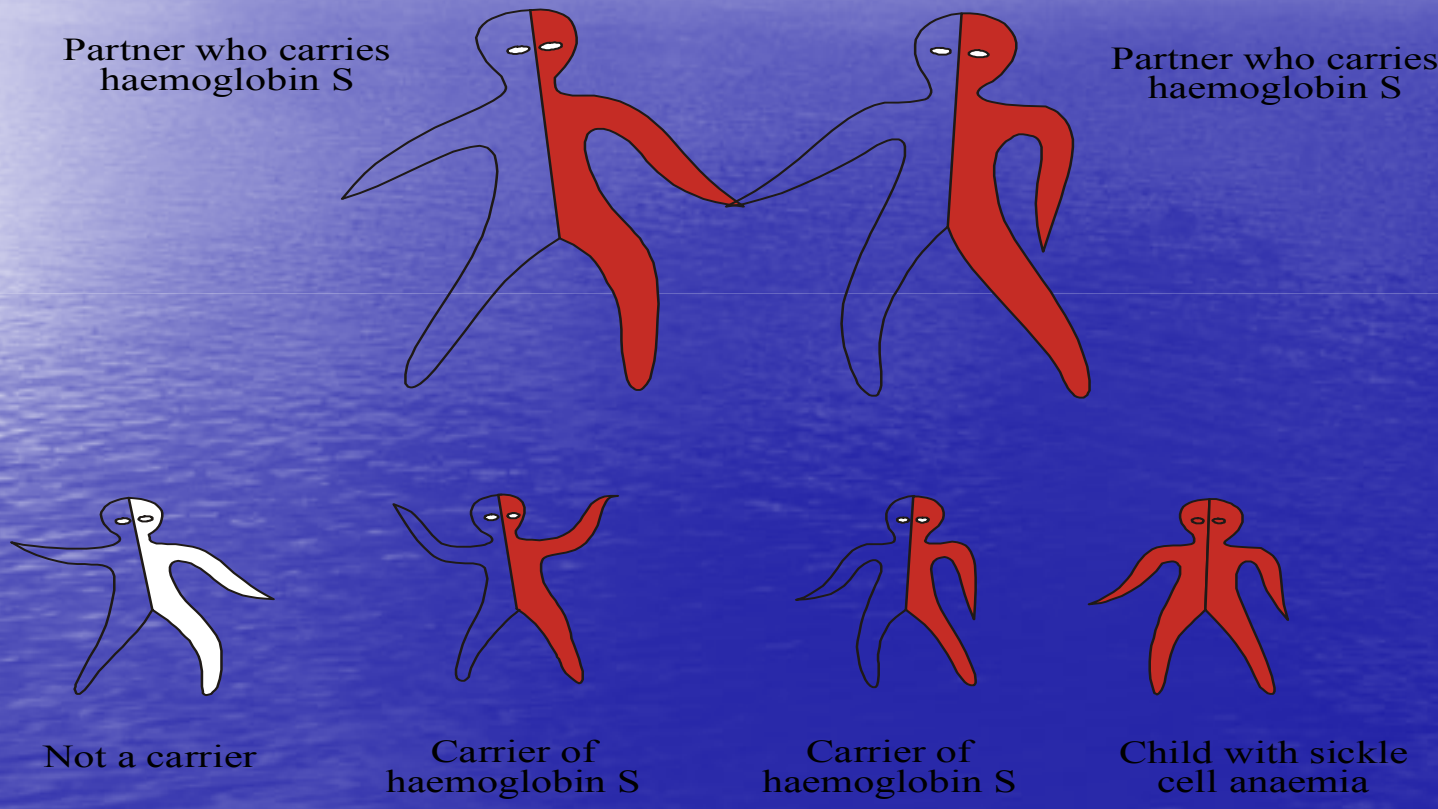
# Forms of Thalassaemia

- Thalassaemia Trait/Minor: people with Thalassaemia trait are perfectly healthy themselves but they can pass THALASSAEMIA MAJOR on to their children. They are sometimes called "healthy carriers of Thalassaemia".
- Thalassaemia Major: this is a very serious blood disorder which begins in early childhood. Children who have Thalassaemia major cannot make enough haemoglobin in their blood. They need frequent blood transfusion and medical treatment.

# Who Is At Risk?



# Pattern of inheritance of SCD and Thalassaemia



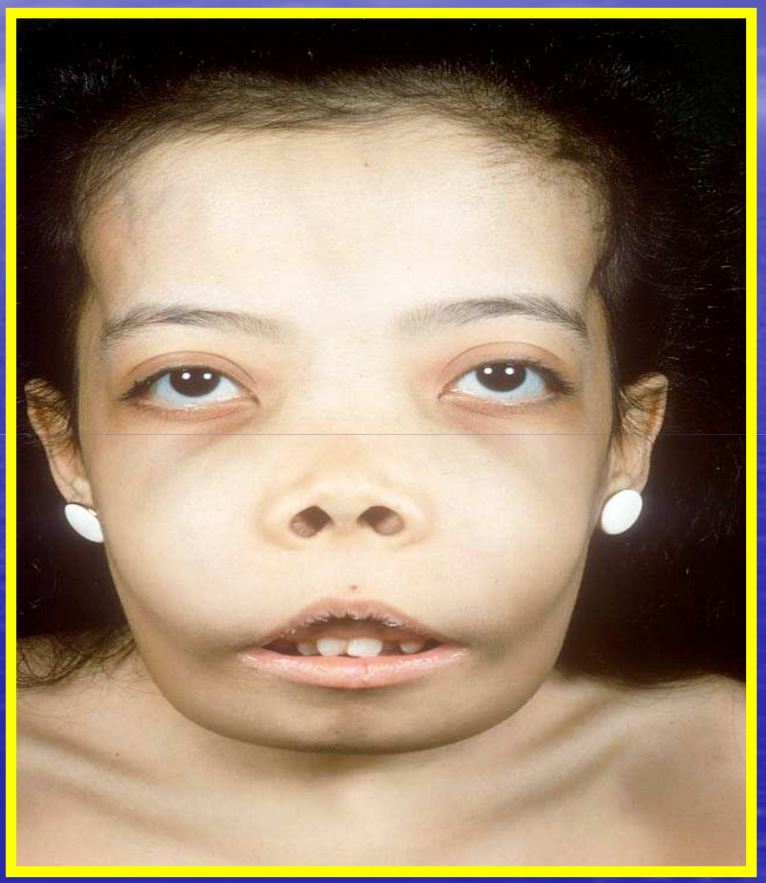
# Symptoms of Thalassaemia

- Paleness
- Fatigue/weakness
- Yellow colored skin (Jaundice)
- Failure to grow & thrive
- Shortness of breath
- Bony abnormalities
- Enlarged spleen and liver

Children born with Thalassaemia major are normal at birth, but begin to show symptoms from the age of 3 - 18 months.

If not treated they usually die between 1 – 8 years old.

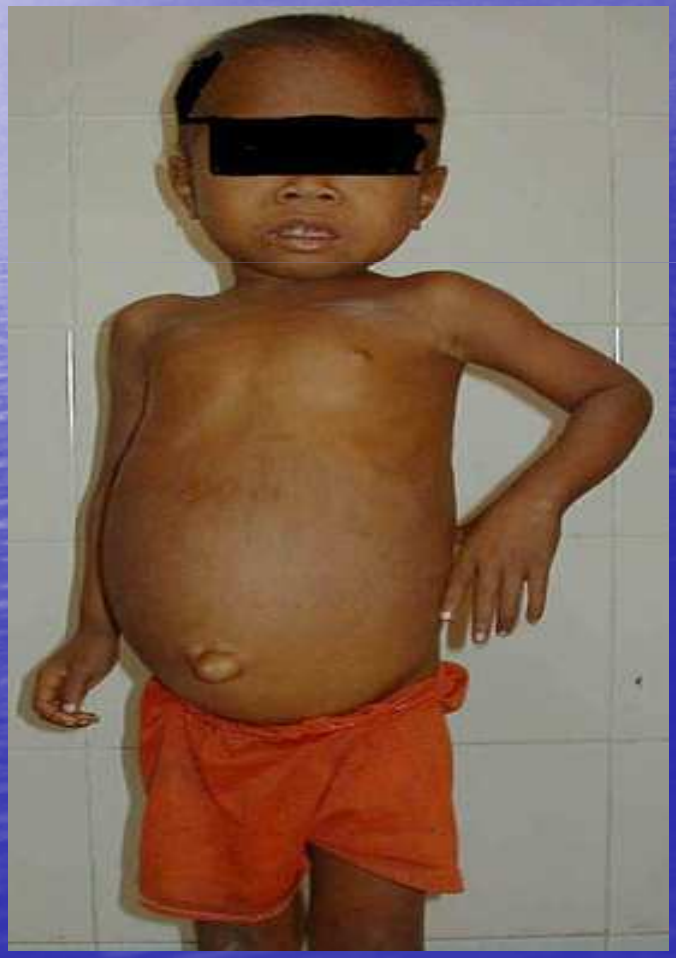
# Symptoms of Thalassaemia

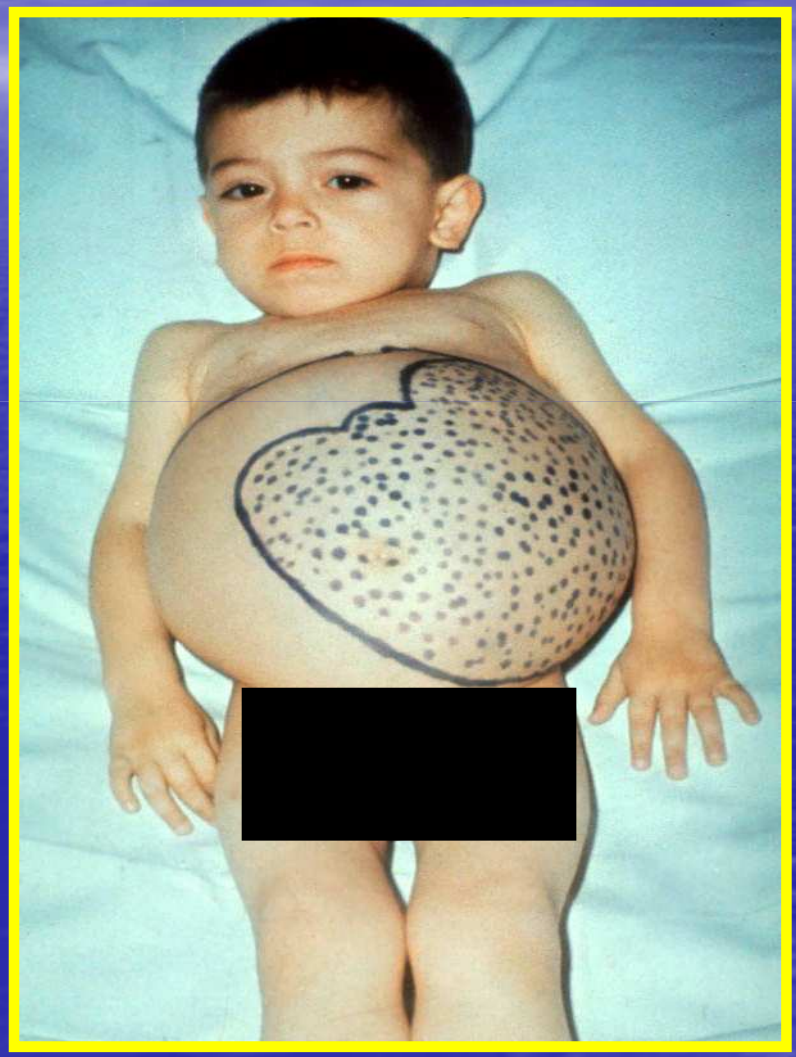


Bony abnormalities (especially of the facial bones)

# Symptoms of Thalassaemia

**Enlarged spleen and liver**



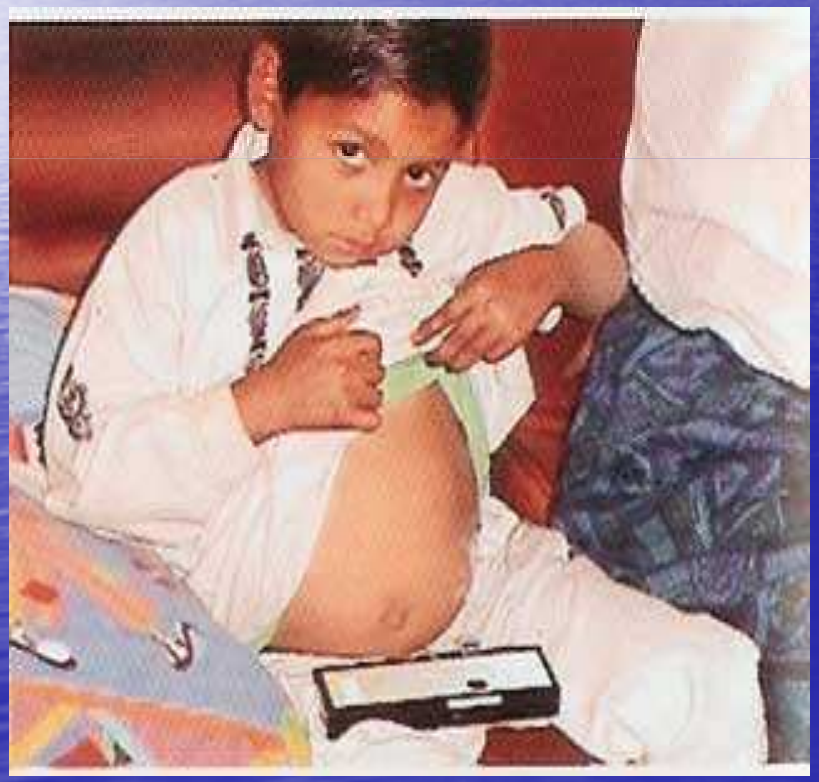


# How is Thalassaemia Managed?

- Regular blood transfusion is required every month throughout life because the body isn't able to make the required amount of haemoglobin.

The blood is imperative for growth, development and a better life.

Due to blood the transfusions, therapy is required to remove the build up of excess iron. This is known as "Iron Chelation".
Defferal infuser is a machine used every 5 - 7 nights a week to get rid of the excess iron.



# Thalassaemia major life expectancy

- Without regular transfusion
  - Less than 10 years
- With regular transfusion and no/poor iron chelation
  - Less than 25 years
- With regular transfusion and good iron chelation
  - ??40 years, ?longer??

# Prevention of Sickle Cell and Thalassaemia

# SCREENING

# Any Questions

# Success Stories

**Dr Ade Olujohungbe**

**Tboz from TLC**

# Success Stories

Pete Sampras

Zinedine Zidane

# AIMS AND OBJECTIVES OF OSCAR SANDWELL CO LTD

- To promote research into the prevention, cure and treatment of Sickle Cell and Thalassaemia.
- To promote public and professional awareness of Sickle Cell and Thalassaemia.
- To advise and support affected families

# OSCAR SANDWELL SERVICES

- Advice & Information
- Support & Advocacy
- Health promotion events
- Workshops &Training
- Screening programmes
- Social events
- Counselling
- Support group
- Reference Library
- Volunteering programmes
- Student Placements

# For further information contact:

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