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SICKLE CELL ANEMIA

INTRODUCTION

Sickle cell anemia is a group of Disease that cause red blood cell to become misshapen and breakdown. A disorder which is inherited from a person's parent. It results in abnormality in the oxygen carrying protein Haemoglobin found in red blood cell. This leads to rigid, sickle like shaped cells.

SYMPTOMS

Attacks of pain
Anaemia
Swelling in hands and feet
Stroke

COMPLICATIONS

Stroke
High blood pressure
Skin ulcers
Gallstones
Blindness
Acute chest syndrome

ETIOLOGY

Genetic cause
The main cause of sickle cell anemia is genetic inheritance.
It occurs when a normal person inherits two abnormal copies of beta globin Gene that makes Haemoglobin.
Normally, humans have Haemoglobin A, Haemoglobin A2, Haemoglobin F in Haemoglobin.
Haemoglobin A which consists of two alpha chains and two beta chains.
In sickle cell anemia both beta chains subunits are replaced with Haemoglobin S in the Haemoglobin.

PATHOPHYSIOLOGY

Normally, RBC are quite elastic and are biconcave disc shape which allows the cells to pass through the capillaries.
In sickle cell anemia,
Inheritance or replacement of beta chains with Haemoglobin S
That lower oxygen tension or Oxygen carrying capacity in the blood
This promotes red blood cell sickling.

Sickling damages the cell membrane and decrease the cells elasticity
Deformity of red blood cell or sickling of cell leads to haemolysis of cell.
Life span of healthy RBC: 90-120 days.
Life span of sickle cell: 10-20 days.

DIAGNOSIS

Diagnosed by:
Sickle solubility test
Haemoglobin with sodium dithionite gives turbid appearance in presence of sickle cell.
If there is no turbid appearance: abscence of sickle cell.