
Good Afternoon
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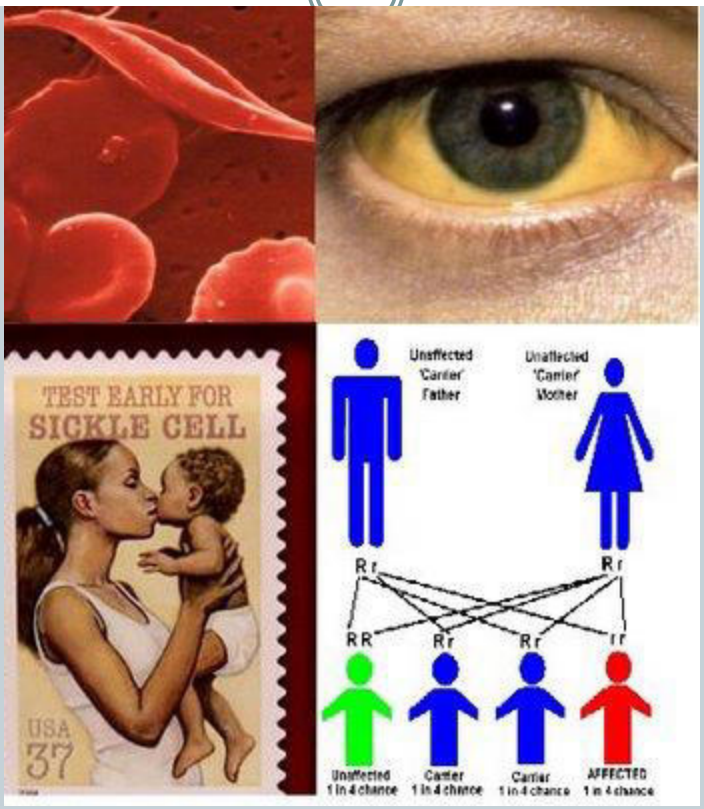
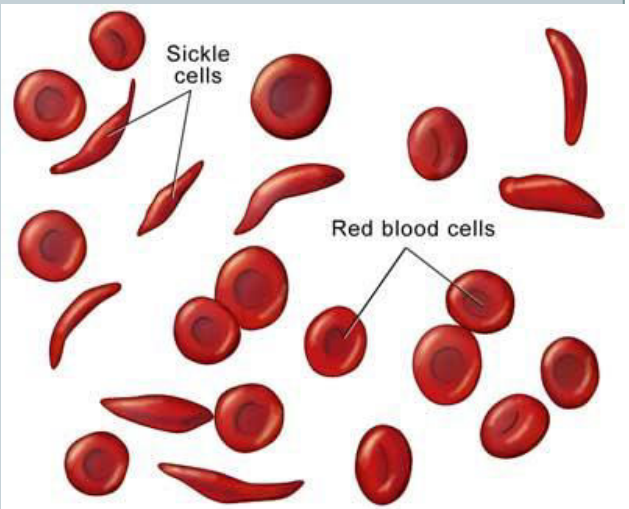
INTRODUCTION

SICKLE CELL ANEMIA



Definition

- A serious condition in which red blood cells can become sickle-shaped due to the presence of abnormal form of hemoglobin ; Hemoglobin S.
- Normal red blood cells are smooth and round in shape. Hemoglobin S causes the erythrocyte to stiffen and elongate taking on sickle shape which are fragile and prone to rupture and decrease the oxygen carrying capacity of them.



Normal and Sickled Red Blood Cells in Blood Vessels

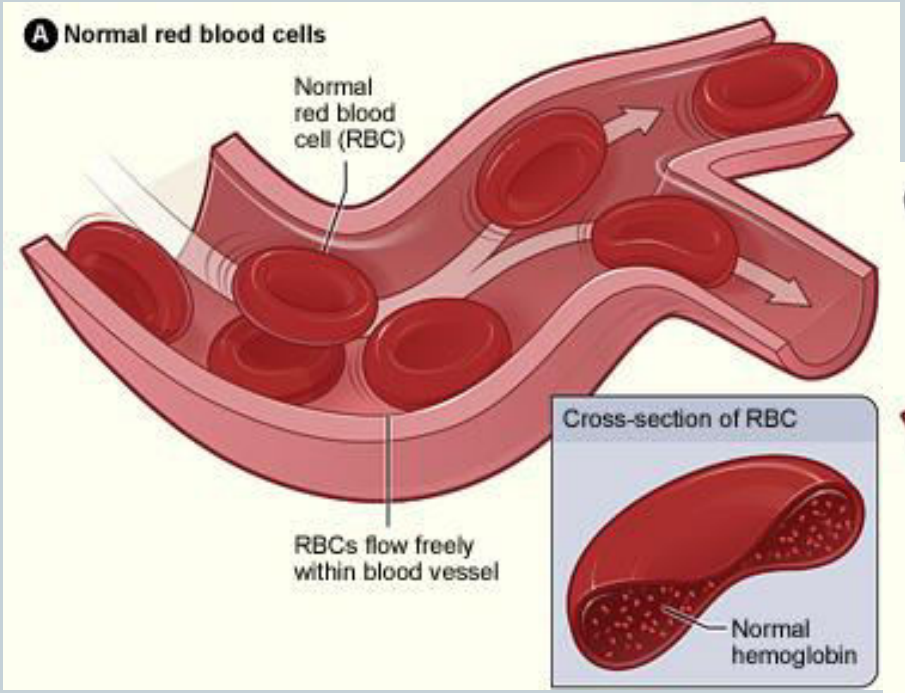
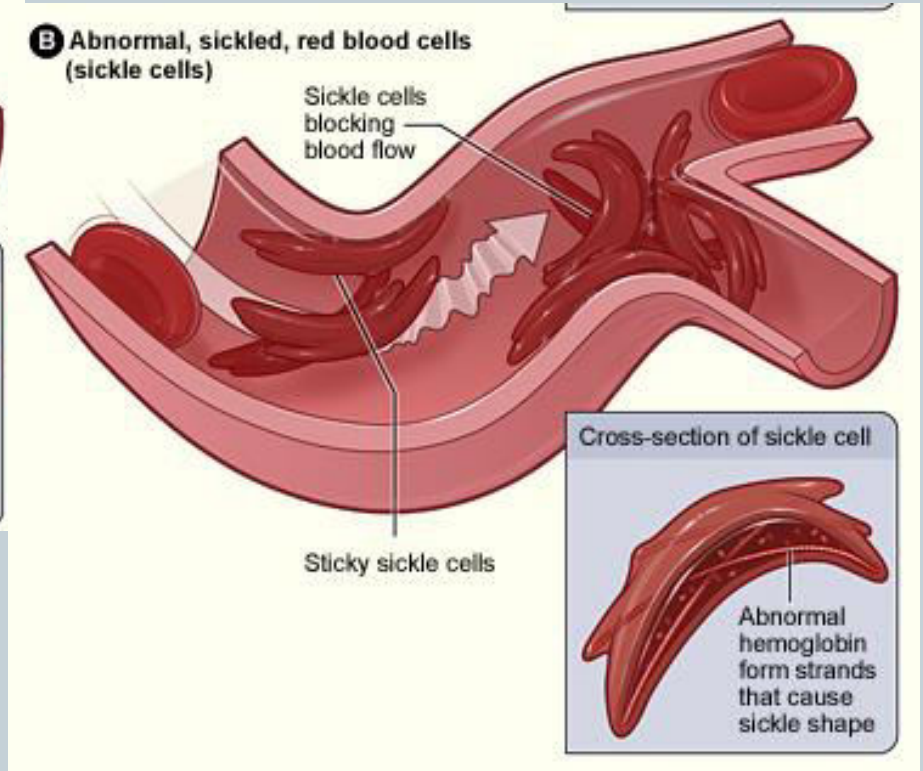


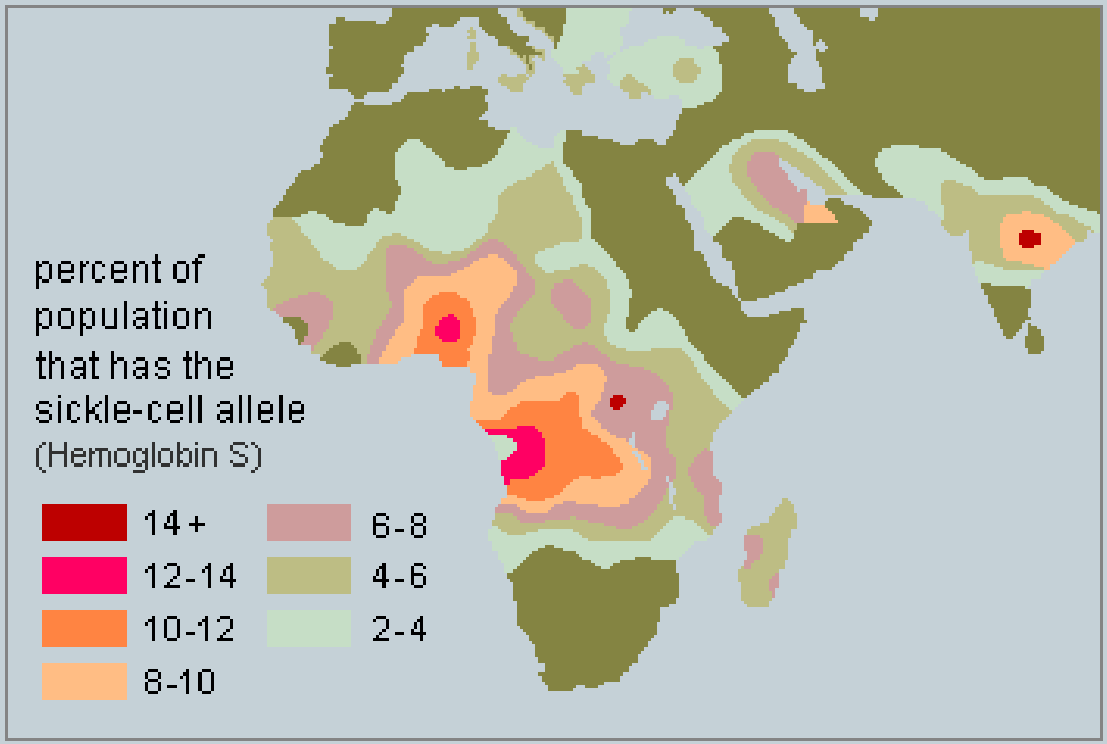
Figure A shows normal red blood cells flowing freely in a blood vessel. The inset image shows a cross-section of a normal red blood cell with normal hemoglobin.

Figure B shows abnormal, sickled red blood cells clumping and blocking the blood flow in a blood vessel. The inset image shows a cross-section of a sickled red blood cell with abnormal strands of hemoglobin.



Source from http://www.nhlbi.nih.gov/health/dci/Diseases/Sca/SCA_WhatIs.html

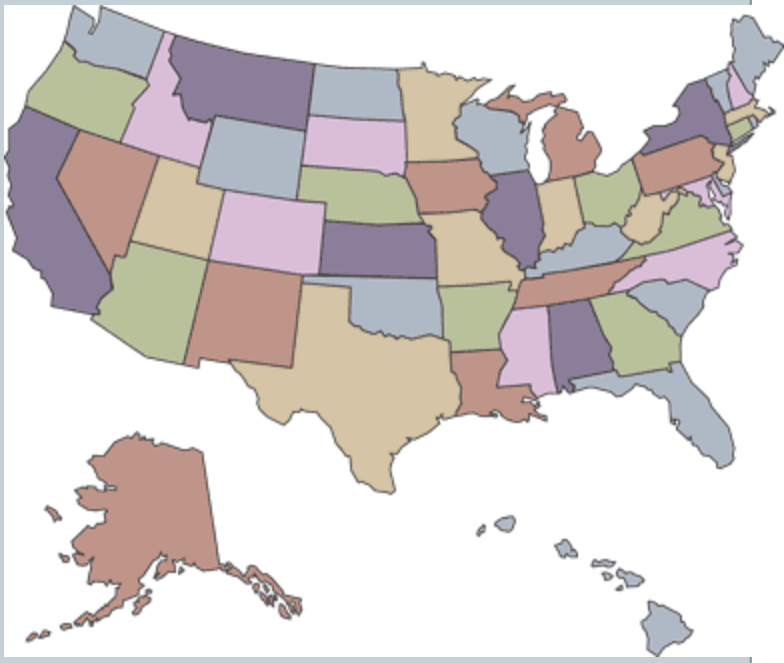
Incidence



- Most common in people whose families come from Africa, South or Central America (especially Panama), Caribbean islands, Mediterranean countries (such as Turkey, Greece, and Italy), India, and Saudi Arabia.

Cont…

- United States, sickle cell anemia affects about 70,000 people.
- Mainly affects African Americans, with the condition occurring in about 1 in every 500 African American births.
- Hispanic Americans also are affected; the condition occurs in 1 out of every 1,000 to 1,400 Hispanic American births.
- About 2 million Americans have sickle cell trait. About 1 in 12 African Americans has sickle cell trait.



Etiology

- Hereditary

Undesirable gene mutation is the main cause of sickle cell anemia

Consagenious marriage increases the incidence rate of sickle cell anemia

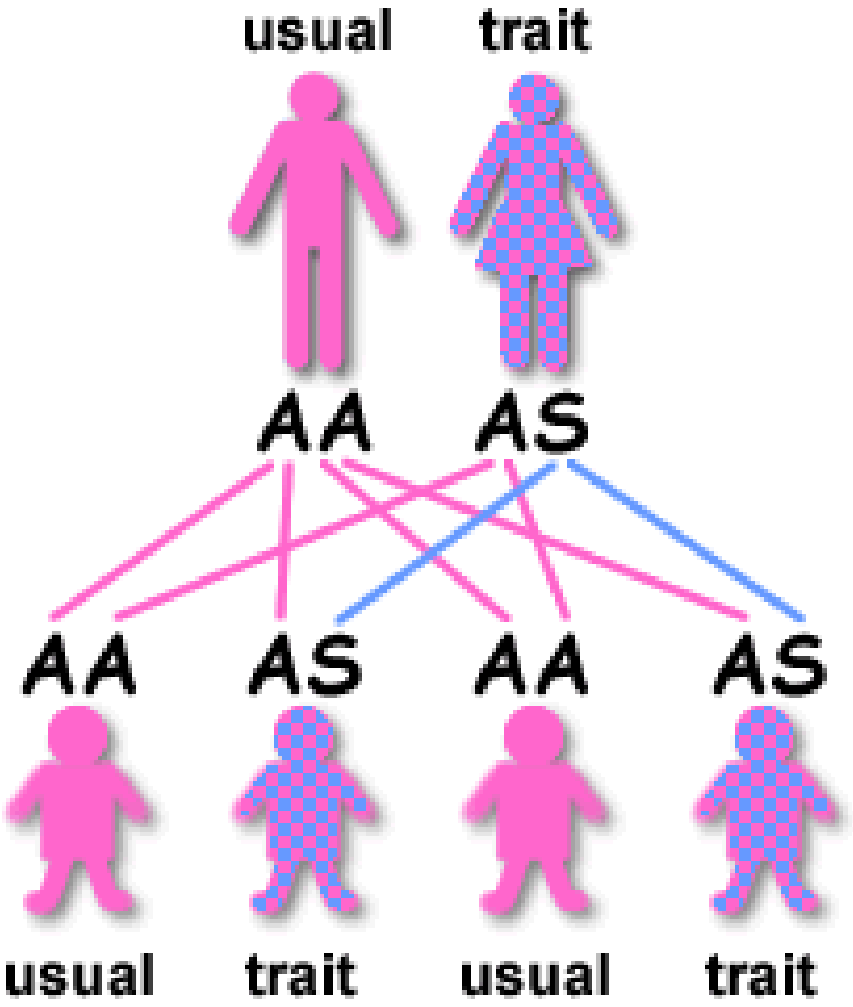
Sickle Cell Anemia vs. Sickle Cell Trait

- People who have sickle cell anemia are born with it; means inherited, lifelong condition.
- They inherit two copies of sickle cell gene, one from each parent.
- Sickle cell trait is different from sickle cell anemia. People with sickle cell trait don't have the condition, but they have one of the genes that cause the condition.
- People with sickle cell anemia and sickle cell trait can pass the gene on when they have children.

If **one parent has sickle cell trait (HbAS) and the other does not carry the sickle hemoglobin at all (HbAA)** then none of the children will have sickle cell anemia.

There is a **one in two** (50%) chance that any given child will get one copy of the HbAS gene and therefore have the sickle cell trait.

It is equally likely that any given child will get two HbAA genes and be completely unaffected.

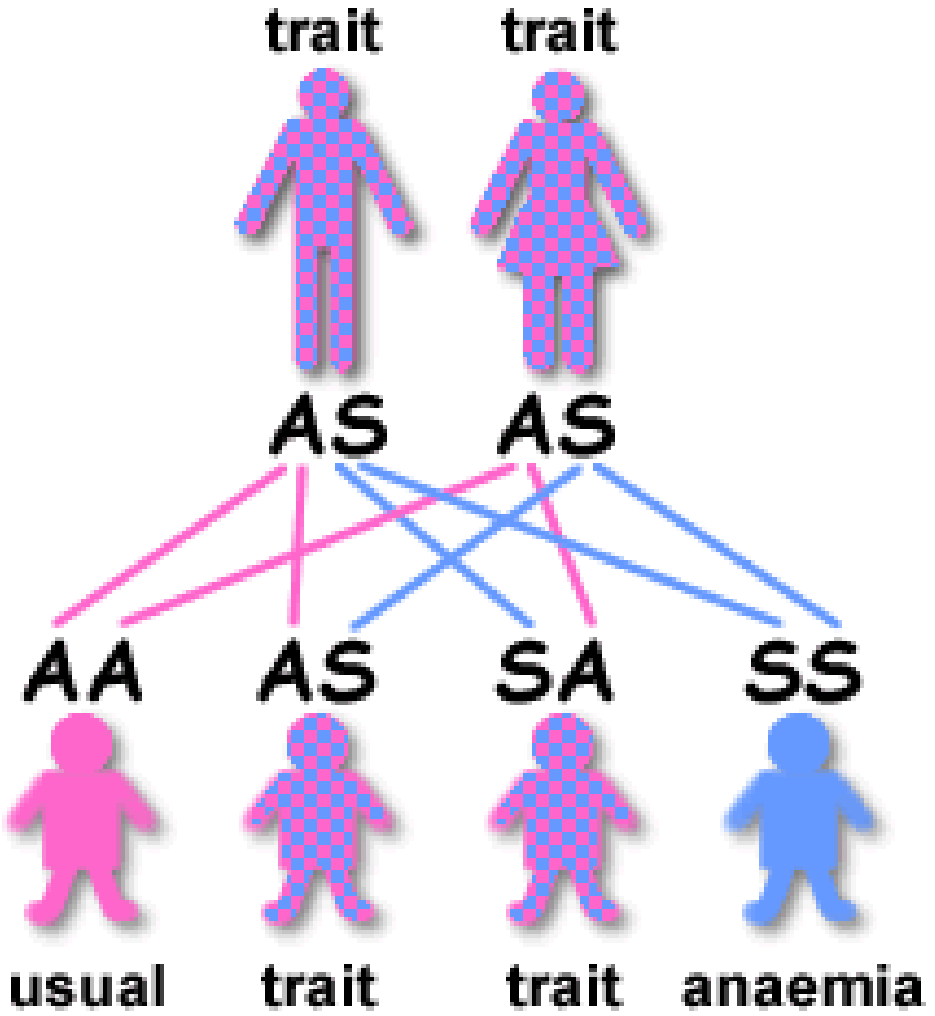


Inheritance of Sickle Cell Anemia

If **both parents have sickle cell trait (HbAS)** there is a **one in four** (25%) chance that any given child could be born with sickle cell anemia.

There is also a **one in four** chance that any given child could be completely unaffected.

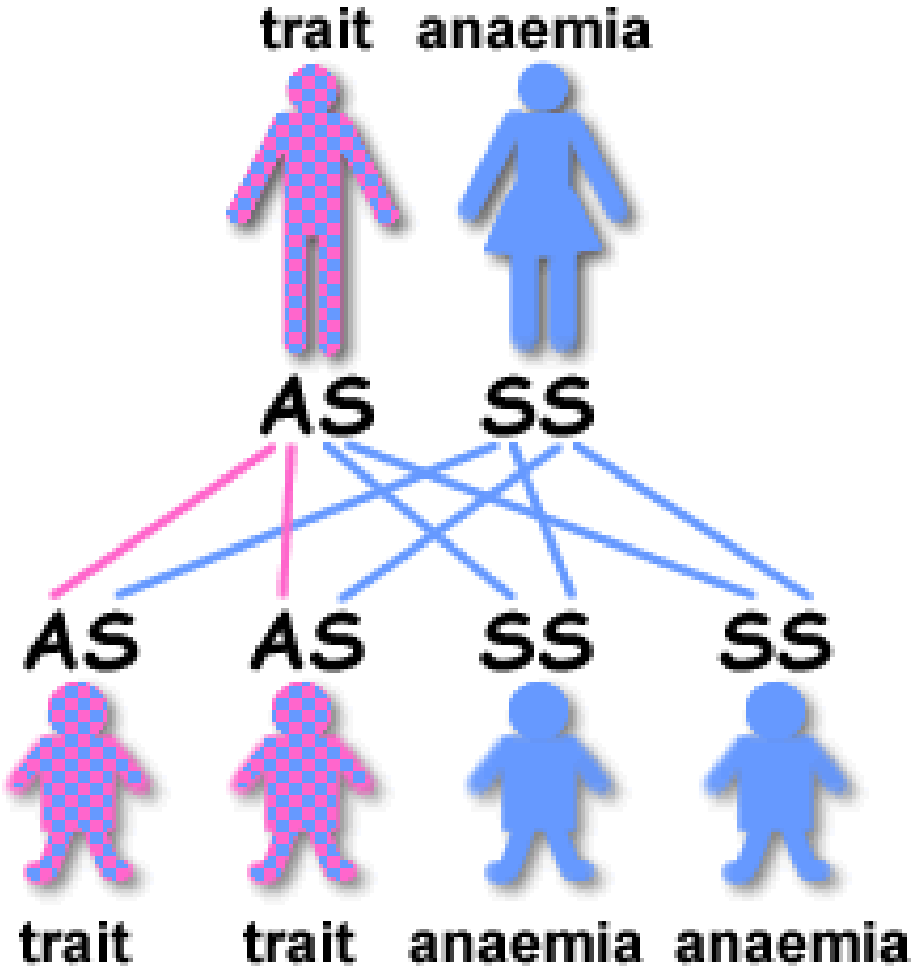
There is a **one in two** (50%) chance that any given child will get the sickle cell trait.



Inheritance of Sickle Cell Anemia

If **one parent has sickle cell trait (HbAS) and the other has sickle cell anaemia (HbSS)** there is a **one in two** (50%) chance that any given child will get sickle cell trait and a **one in two** (50%) chance that any given child will get sickle cell anemia.

No children will be completely unaffected.

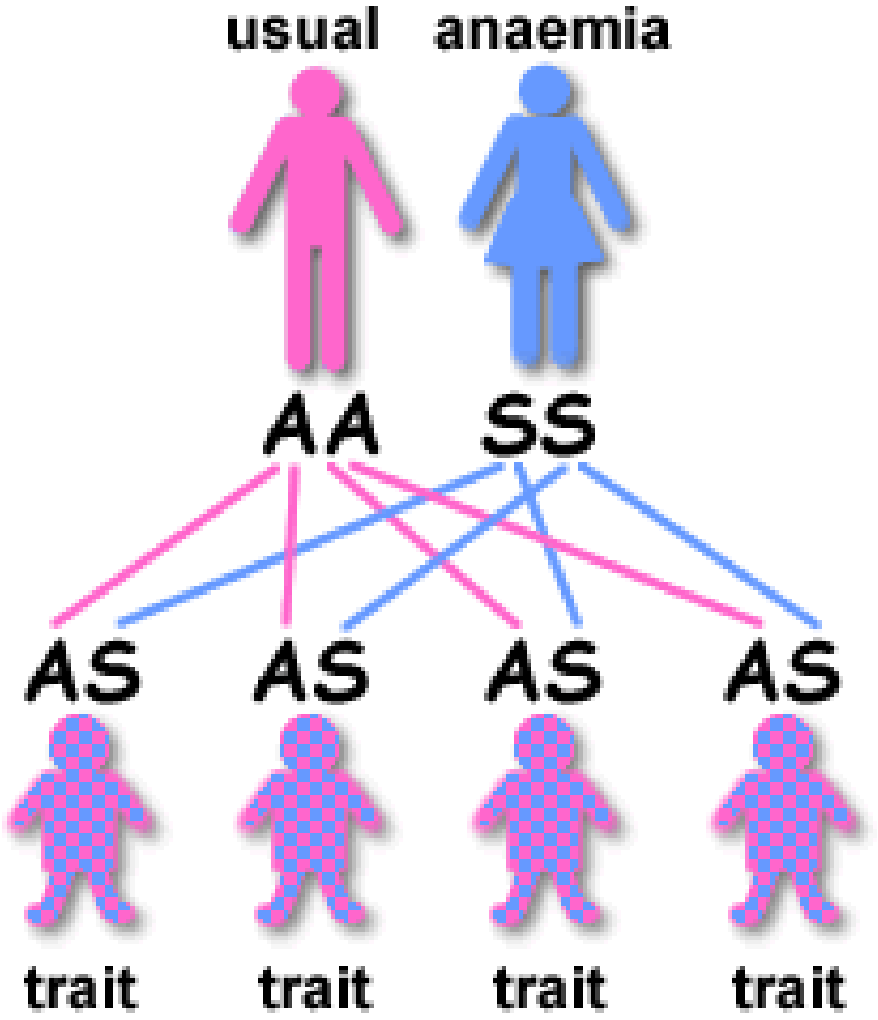


Inheritance of Sickle Cell Anemia

If **one parent has sickle cell anaemia (HbSS) and the other is completely unaffected (HbAA)** then all the children will have sickle cell trait.

None will have sickle cell anemia.

The parent who has sickle cell anemia (HbSS) can only pass the sickle hemoglobin gene to each of their children.



Inheritance of Sickle Cell Anemia

Conditions Trigger Sickling

- Dehydration
- Acidosis
- Low body temperature
- Decreased Plasma volume

Pathophysiology

Due to triggering factors

Sick ling of RBCs

Change in shape, elastiasty & O2 carrying capacity of RBCs

Hemolysis by Spleen

Anemia

Occlusion of small blood vessels

Changes in capillary membrane permeability

Plasma loss &

Hemoconcentration

↓

Thromboses formation

↓

Tissues necrosis

Clinical Manifestations

- Pain ; due to tissue hypoxia and damage
- Fatigue and pallor
- Fever
- Swelling and tenderness
- Tachypnea
- Hypertension
- Nausea and vomiting
- Dactylitis – Swelling and inflammation of hands and feet
- Lug ulcers

Diagnostic Evaluations

- History collection and physical examination
- Complete blood count
- Peripheral blood smear
- Serum bilirubin – increased due to hemolysis
- D. N.A testing – rarely done
- Doplar studies – to detect deep vein thrombosis
- X- ray , M.R.I & C.T. scans

Cont....

- Sickle solubility test :- A mixture of hemoglobin S in reducing solution such as sodium dithionite gives a turbid appearance where as the normal Hb gives a clear solution

- Sickling Test :- RBCs are exposed to a deoxygenating agent

Complications

- Brain :- thrombosis , hemorrhage
- Lung :- Acute chest syndrome , Pulmonary hypertension , Pneumonia
- Heart :- Heart failure
- Spleen :- Splenic atropy
- Kidney :- Hematuria and renal failure
- Liver and Gall bladder :- Hepatomegali & gall stones

Cont....

- Bones and Joints :- Hand and foot syndrome

Osteonecrosis , Osteolysis , Bone shortening

- Skin :- Ulcers of hands , ankles and feet
- Infection :- Several infections including Pneumonia , bronchitis, cystitis , meningitis , sepsis etc.

Management

There is no specific pharmacological or surgical management for sickle cell anemia

Pharmacological management include

- Symptomatic treatment
- Pain management
- Prevention of complications

Pharmacological Management

- Transfusion therapy :- Long term RBC transfusion
- Pain Management :-
  I. Opioids
  II. NSAIDs
  III. Local anesthetics

O2 Therapy

Fluid and electrolyte replacement

Folic acid supplimentation

Stem Cell transplantation

- Autologus Transplant
- Allogenic Transplant

Collection Process of Stem cells

1. Apherisis :- By machiene
2. Directly from Pelvis of the Donor – Rare procedure.
3. From umbilical Cord and Placenta

Nursing Managemet

Assessment

- Pain assessment
- Vital signs
- Complications
- Neurological assessment
- Integumentary assessment

Nursing Diagnosis

- Acute pain relate to tissue hypoxia secondary to occlusion of blood vessels by sickled cells
- Risk for infection
- Activity intolerance related to powerlessness secondary to decreased blood supply
- Knowledge deficit regarding the disease condition

Nursing Interventions

Prognosis

Prognosis is guarded for sickle cell anemia

The goal for treatment is to achieve normal life span with minimal morbidity. As therapy improves prognosis also improves

More often the patients with sickle cell anemia dies due to secondary causes. The leading cause of death are acute chest syndrome , pulmonary embolism and infection.


CONCLUSIÓN

Assignment

Write a nursing care plan for the patient with sickle cell anemia

Date for submission :- 23 / 01 / 2012

Reference

Suddarth and Brunner , Text book of medical and surgical nursing , 10th Edition , Lippincott Williams and Wilkins , Philadelphia , 2008


Thank
You…