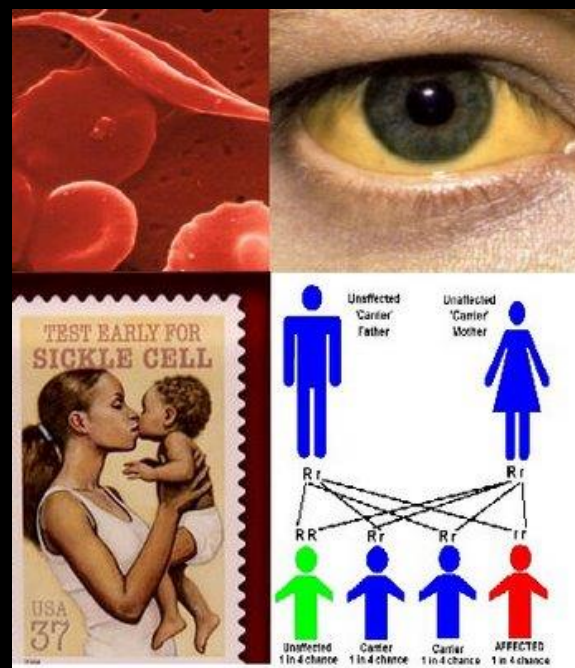


SICKLE CELL ANEMIA



Sickle cell disease is hereditary hemoglobinopathy, characterized by the partial or complete replacement of normal hemoglobin with abnormal hemoglobin S (Hgb S) in red blood cells. Sickle cell trait (Hgb S

Sickle cell anemia is the most common type of Sickle cell disease are

1-Sickle cell Trait (Hgb SA)

Heterozygous condition(child has one sickle cell hemoglobin gene and one normal hemoglobin gene

2-1-Sickle cell anemia (Hgb SS)

Homozygous condition(child has two sickle cell hemoglobin gene

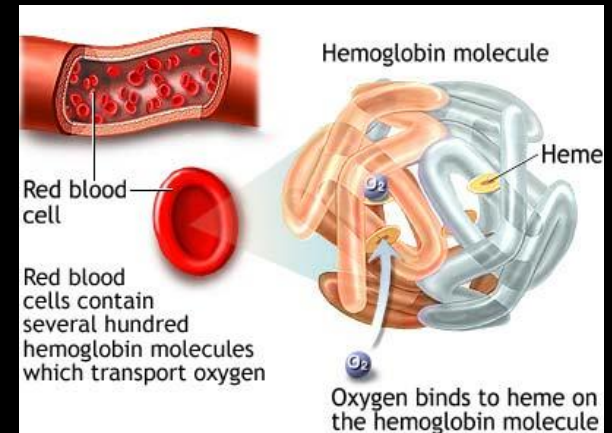
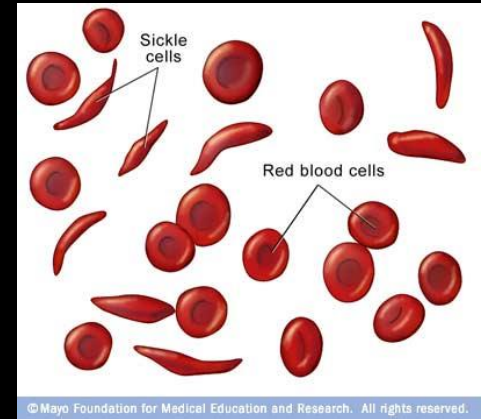
3-1-Sickle cell Syndrome

C shape instead of an S shape

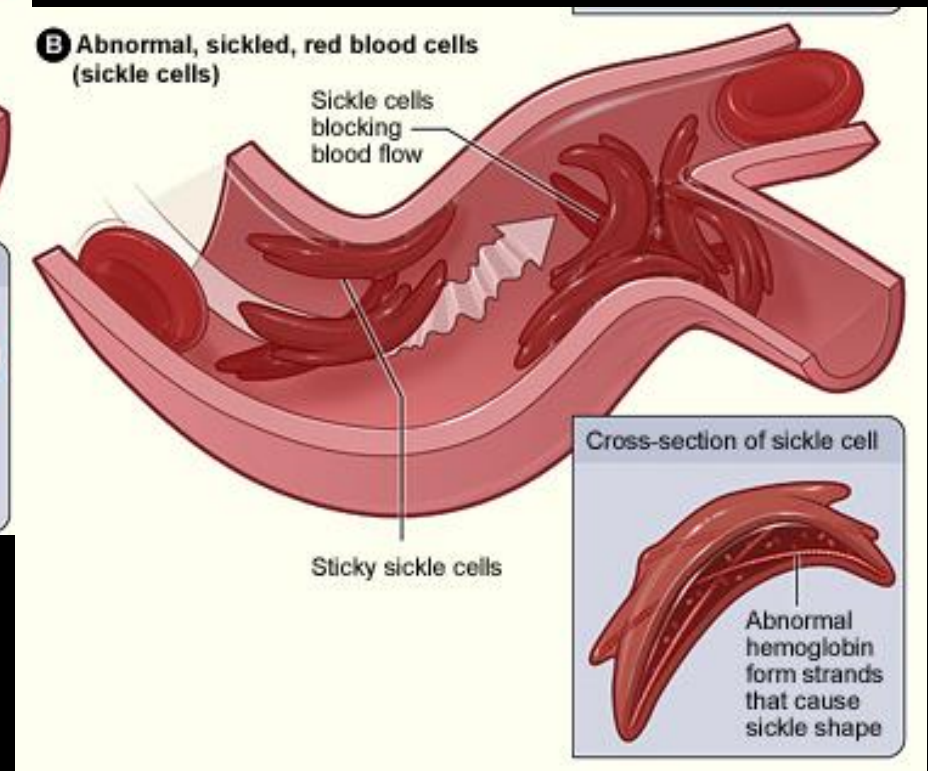
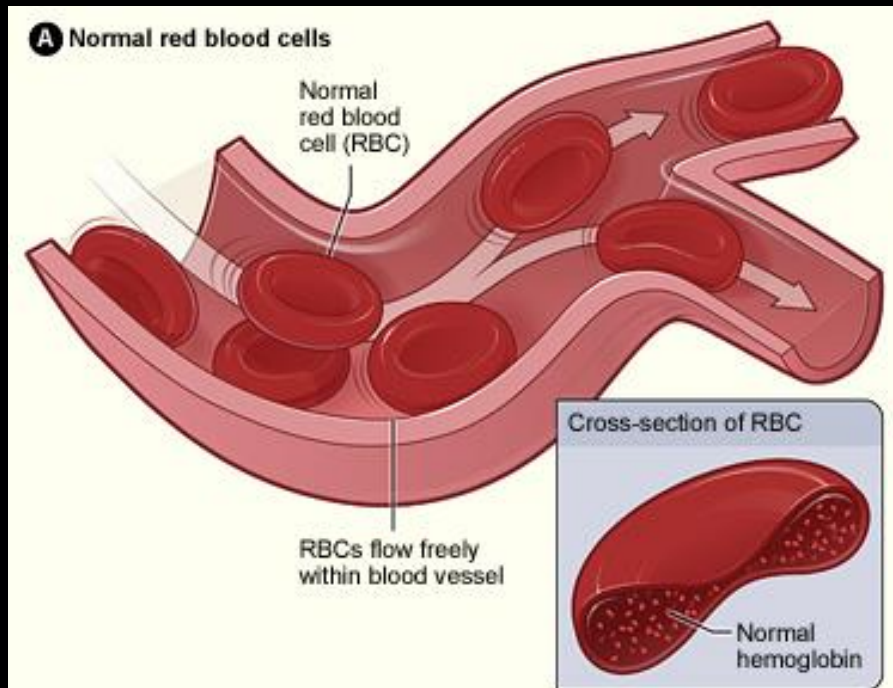
4-1-Sickle cell-B-Thalassemia Disease (Hgb SB)

What is Sickle Cell Anemia?

- A serious condition in which red blood cells can become sickle-shaped
- Normal red blood cells are smooth and round. They move easily through blood vessels to carry oxygen to all parts of the body.
- Sickle-shaped cells don't move easily through blood. They're stiff and sticky and tend to form clumps and get stuck in blood vessels.
- The clumps of sickle cell block blood flow in the blood vessels that lead to the limbs and organs. Blocked blood vessel can cause pain, serious infection, and organ damage.



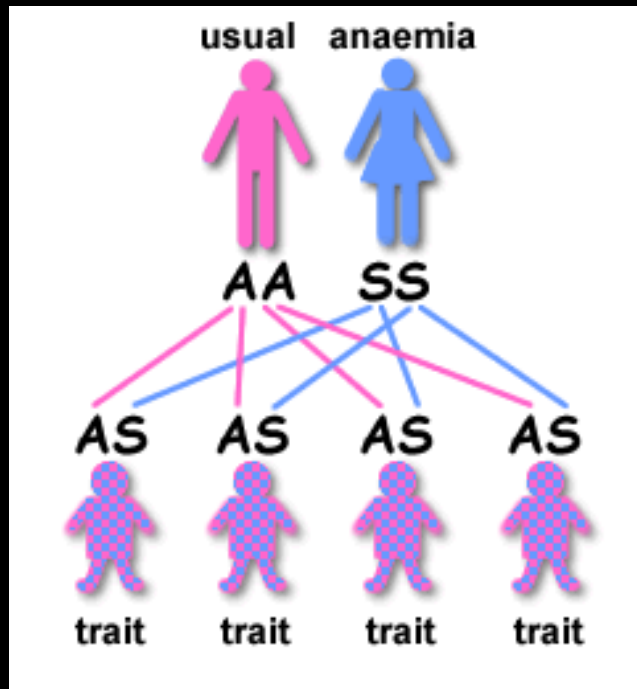
Normal and Sickled Red Blood Cells in Blood Vessels



Sickle Cell Anemia vs. Sickle Cell Trait

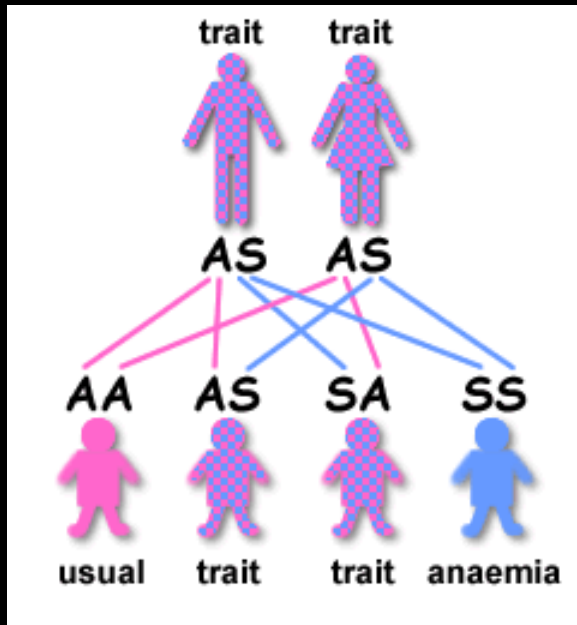
- People who have sickle cell anemia are born with it; means inherited, lifelong condition.
- They inherit two copies of sickle cell gene, one from each parent.
- Sickle cell trait is different from sickle cell anemia. People with sickle cell trait don't have the condition, but they have one of the genes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

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.

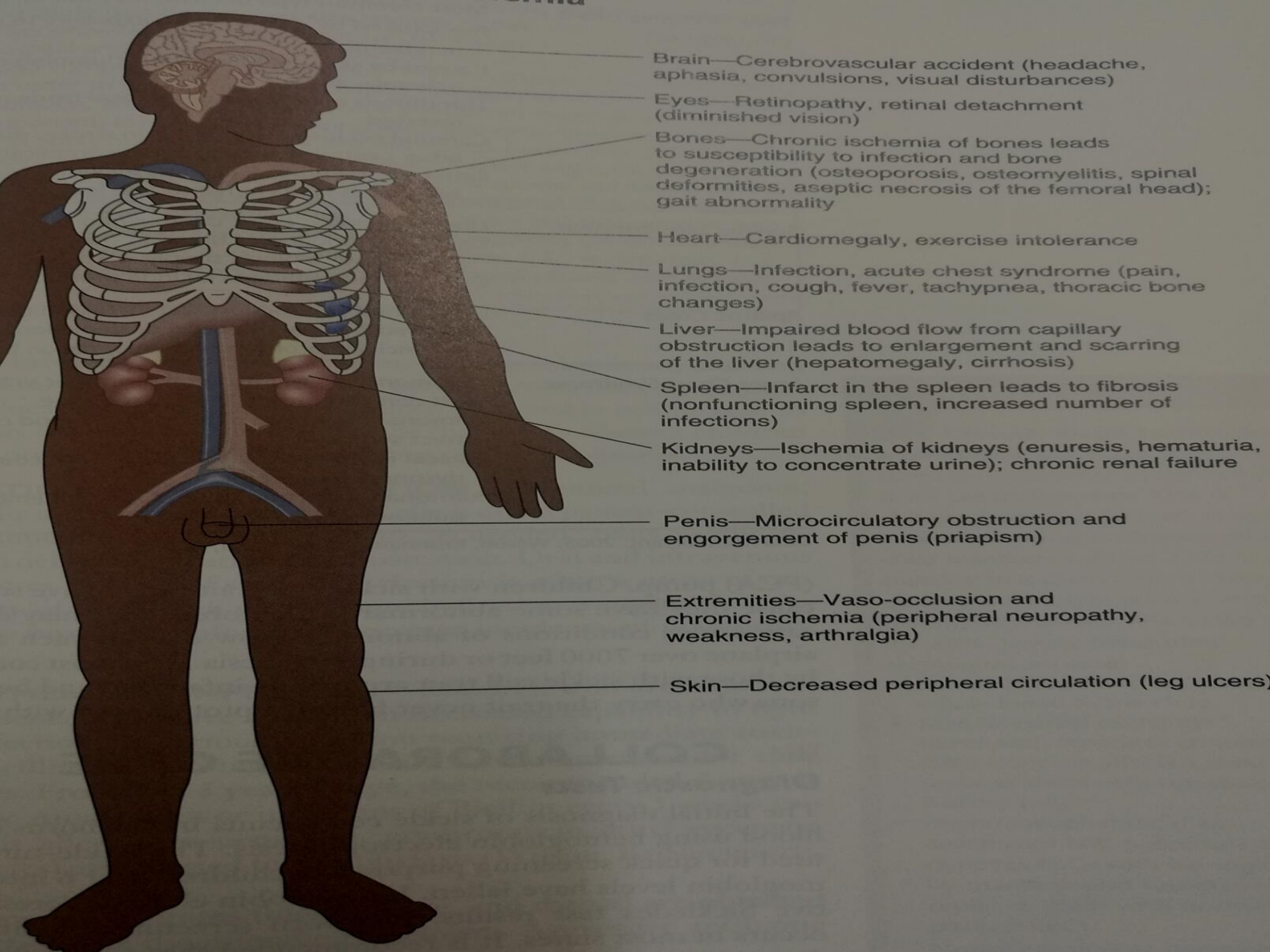
Inheritance of Sickle Cell Anemia



- 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.

Why Anemia?

- [Anemia](#) is a condition in which a person's blood has a lower than normal number of red blood cells, or the red blood cells don't have enough hemoglobin. Hemoglobin is an iron-rich protein that gives blood its red color and carries oxygen from the lungs to the rest of the body.
- Normal red blood cells last about 120 days in the bloodstream and then die. Their main role is to carry oxygen, but they also remove carbon dioxide (a waste product) from cells and carry it to the lungs to be exhaled.
- In sickle cell anemia, a lower-than-normal number of red blood cells occurs because sickle cells don't last very long. Sickle cells die faster than normal red blood cells, usually after only about 10 to 20 days. The bone marrow can't make new red blood cells fast enough to replace the dying ones. The result is anemia.



Brain—Cerebrovascular accident (headache, aphasia, convulsions, visual disturbances)

Eyes—Retinopathy, retinal detachment (diminished vision)

Bones—Chronic ischemia of bones leads to susceptibility to infection and bone degeneration (osteoporosis, osteomyelitis, spinal deformities, aseptic necrosis of the femoral head); gait abnormality

Heart—Cardiomegaly, exercise intolerance

Lungs—Infection, acute chest syndrome (pain, infection, cough, fever, tachypnea, thoracic bone changes)

Liver—Impaired blood flow from capillary obstruction leads to enlargement and scarring of the liver (hepatomegaly, cirrhosis)

Spleen—Infarct in the spleen leads to fibrosis (nonfunctioning spleen, increased number of infections)

Kidneys—Ischemia of kidneys (enuresis, hematuria, inability to concentrate urine); chronic renal failure

Penis—Microcirculatory obstruction and engorgement of penis (priapism)

Extremities—Vaso-occlusion and chronic ischemia (peripheral neuropathy, weakness, arthralgia)

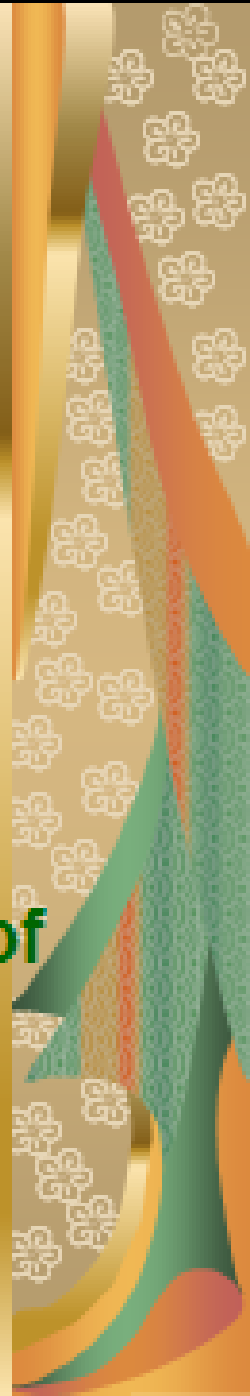
Skin—Decreased peripheral circulation (leg ulcers)

Complication of Sickle Cell Anemia

- Hand-Food Syndrome
- Splenic Crisis
- Infections
- Acute Chest Syndrome
- Delayed growth and puberty in children
- Stroke
- Eye problem
- Priapism
- Gallstone
- Ulcers on the legs
- Pulmonary Arterial Hypertension
- Multiple Organ Failure

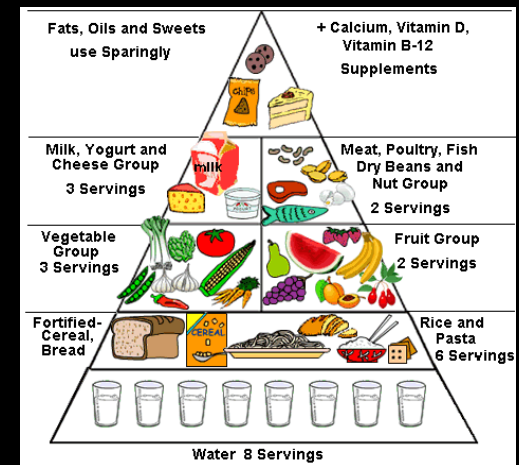
Treatment

1. folic acid
2. Analgesics
3. **BMT**
4. **Antibiotics**
5. **Additional treatments may include:**
 - *Dialysis or kidney transplant
 - *Surgery for eye problems
 - *Hip replacement for a vascular necrosis of the hip
 - *surgery for leg ulcers
 - *counseling for the psychosocial complications



Prevention

- Identify what can trigger the “Crisis” such as stress, avoid extremes of heat and cold weather,
- Maintain healthy lifestyle habits
 - ✦ Eating healthy
 - ✦ Avoid dehydration
 - ✦ Exercise regularly
 - ✦ Get enough sleep and rest
 - ✦ Avoid alcohol and don’t smoke
- Regular medical checkups and treatment are important



Nursing management:

A- Prevent sickling by:

⊠ **promote tissue oxygenation and prevent tissue hypoxia through:**

- Avoid Strenuous physical activity
- Emotional stress
- Avoid Environments with low oxygen content

⊠ **promote proper hydration by:**

- Recognize signs of dehydration and Provide access to fluids
- Avoid excess exposure to the sun

⊠ **prevent infection by:**

- Keep child properly immunized as recommended by the health care provider
- Early detection of signs and symptoms of infection
- Frequent measured of vital signs

B- Promote supportive therapies during crises

⊠ **Control pain by:**

- Administer analgesic and Comfortable position
- apply warmth to painful area

⊠ **promote rest by keep child at bed rest**

C-Replace blood and observe for signs of transfusion reactions

D- Prevent psychological problem

E-Promote long term care and genetic counseling