

*Tikrit University*

*College of Nursing*

*Clinical Nursing Sciences*



**Third Year - 2023-2024**

**Child Health Nursing**



**Sickle cell anemia (SCA) (Hemoglobinopathies)**

*by:*

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## **Sickle cell anemia (SCA) (hemoglobinopathies)**

In which normal adult Hgb [HbA] is partly or completely replaced by abnormal sickle Hgb (**HbS**).

Single base mutation in the DNA (**Valine** instead of **Glutamic acid**) in position number 6 of beta chains of the hemoglobin Molecule

### **Clinical Manifestations of Sickle Cell Anemia**

#### **General**

1. Growth retardation
2. Chronic anemia (hemoglobin level of 6–9 g/dl)
3. Delayed sexual maturation
4. Marked susceptibility to sepsis

#### **Vasoocclusive Crisis**

Pain in area(s) of involvement (Ischemia of involved areas)

**Extremities**—Painful swelling of hands and feet (sickle cell dactylitis, or hand–foot syndrome), painful joints

**Abdomen**—Severe pain resembling acute surgical condition

**Cerebrum**—Stroke, visual disturbances

**Chest**—Symptoms resembling pneumonia, protracted episodes of pulmonary disease

**Liver**—Obstructive jaundice, hepatic coma

**Kidney**—Hematuria

**Genitalia**—Priapism (painful penile erection)

#### **Sequestration Crisis Nursing Care Management**

1. Family & patient Health (SCA) Education
2. Keep adequate hydration to prevent sickling and to delay **adhesion–stasis–thrombosis–ischemia cycle**.
3. Not sufficient to advise parents to “force fluids” or “encourage drinking.”
4. Specific instructions (how many daily glasses or bottles of fluid are required.
5. Avoid limiting fluids at night.
6. Enuresis is treated as a **complication of the disease**, like joint pain
7. Alleviate parental pressure on the child.
8. Pain program should be combined with **psychological support**
9. In case of blood transfusions or exchange transfusions, the nurse (**observe for signs of transfusion reaction**).
10. Avoid hypervolemia by rapid transfusion because of heart failure
11. Look for signs of cardiac failure.
12. Check splenic size because **increasing splenomegaly** is an ominous sign to avoid splenic sequestration

13. A decreasing spleen size **denotes response to therapy**.
14. Vital signs & BP are also monitored for impending shock.
15. Record intake, (IV fluids), and output.
16. Check weight to serve as a baseline for evaluating **hydration**.
17. **Look for signs of hypokalemia because diuresis**
18. Look for signs of **Acute Chest Syndrome and Cerebrovascular accident** (fatal complication)

### **Therapeutic Management**

1. Treatment of underlying causes of IDA (ancylostma, peptic ulcer)
2. Dietary counseling & oral Iron supplements
3. In formula-fed infants, (iron-fortified commercial formula & iron fortified infant cereal.
4. Infants < 12 months avoid fresh cow's milk because it may increase the risk of GI blood loss
5. If GI bleeding is suspected, child's stool guaiac tested at least 4-5 times to identify any intermittent blood loss.
6. Dietary iron-rich foods is **inadequate** as treatment of IDA (iron is poorly absorbed)
7. Oral iron supplements for 3 months.
8. Ferrous iron, more readily absorbed than ferric iron,
9. Ascorbic acid (vitamin C) facilitate absorption of iron & given as vitamin C-enriched foods and juices with the iron preparation.
10. Response to iron therapy can be monitored by
  - **reticulocyte count**
  - **Hb level** (Hb should rise by 1 gm % per week)
11. Oral: tab/ 8 hrs.
  - Ferrous sulphate 200 mg tab
  - Ferrous gluconate 300 mg tab
12. If Hgb fails to rise after 1 month of oral therapy, assess for
  - persistent bleeding,
  - iron malabsorption,
  - noncompliance,
  - improper iron administration,
13. Parenteral (IV or IM) iron administration is safe and effective but
  - **painful,**
  - expensive,
  - regional lymphadenopathy,

- transient arthralgias
- serious allergic reaction.
- carcinogenic

#### **14. Parental (IM or IV)**

- Iron -Dextran: 100 mg / IM or IV
- Iron Sorbitol: 50 mg / IM

#### **Indication of parental: if**

- oral iron intolerance
- malabsorption syndrome
- GI disorders (peptic ulcer)
- rapid iron loss

#### **Indications of blood Transfusions in**

- Severe anemia
- Serious infection,
- Cardiac dysfunction,
- Surgical emergency when anesthesia is required.

**Packed RBCs (2–3 ml/kg)**, not whole blood, are used to minimize the chance of circulatory overload. Supplemental oxygen is administered when tissue hypoxia is severe

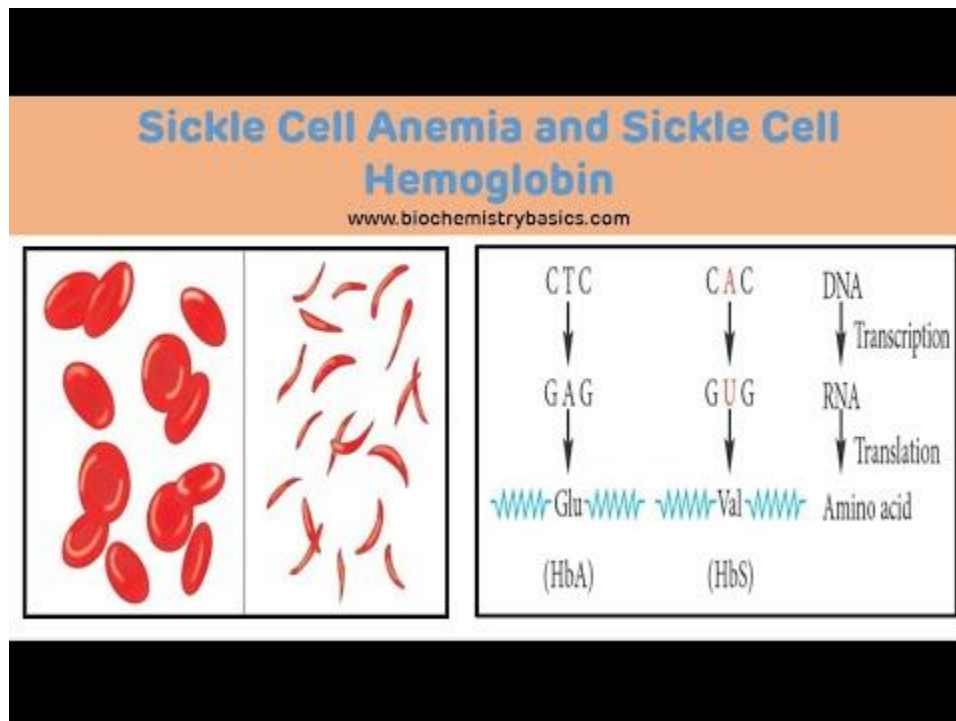
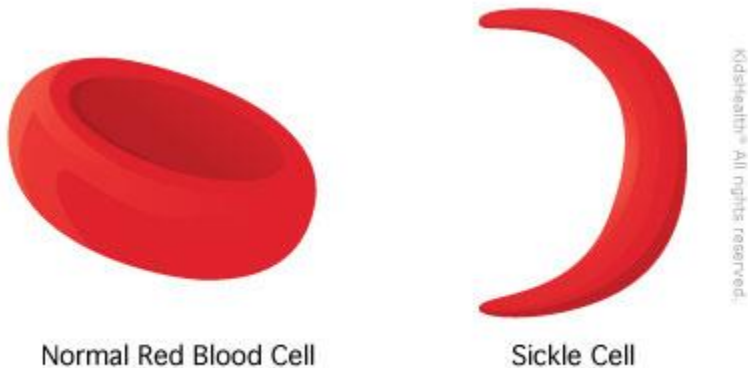
#### **Nursing Care Management**

1. Keep adequate iron intake
2. Educate the parents about Protection from injury due to an unsteady gait or dizziness.
3. Instruction on Iron administration.
4. **Oral iron** given in 2 divided doses between meals.
5. An adequate dosage of oral iron turns the stools a tarry green color, Vomiting or diarrhea can occur with iron therapy.
6. If parenteral iron are prescribed, iron dextran must be injected deeply into a large muscle mass using the Z-track method. The injection site is not massaged after injection to minimize skin staining and irritation. Because no more than 1 ml should be given in one site, the IV route should be considered to **avoid multiple injections**.
7. observe for allergic reactions (anaphylaxis, with IV administration.
8. A test dose is recommended before routine use.

9. In formula-fed infants educate on importance of iron-fortified formula and of **introducing solid foods** at appropriate age during the first year of life.

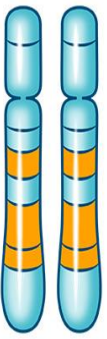
10. Cereals are first semisolid foods to be introduced into infant's diet at 6 months of age.

The nurse can also stress that overweight is not synonymous with good health.




# What is Sickle Cell Disease (SCD)?

**Chromosome 11**




Two copies of a mutated Hemoglobin Subunit Beta (HBB) gene


**Pain crises due to ischemia**



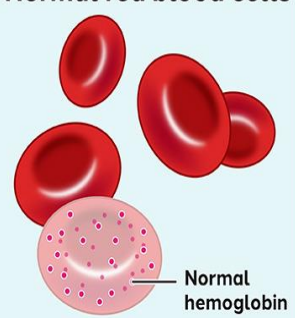
**Anemia**



**Susceptibility to infections**

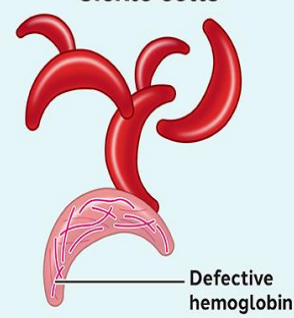


**Normal red blood cells**



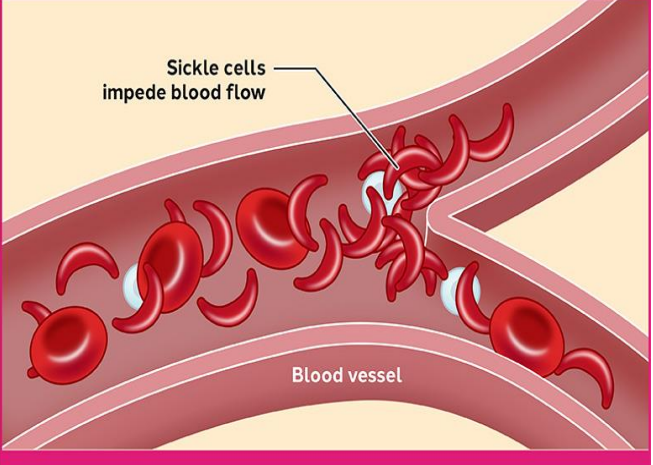
Normal hemoglobin

**Sickle cells**




Defective hemoglobin

**Sickle cells impede blood flow**

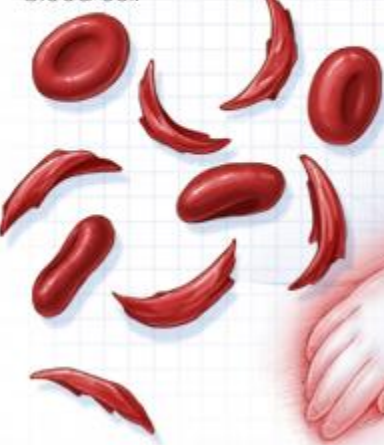


Blood vessel

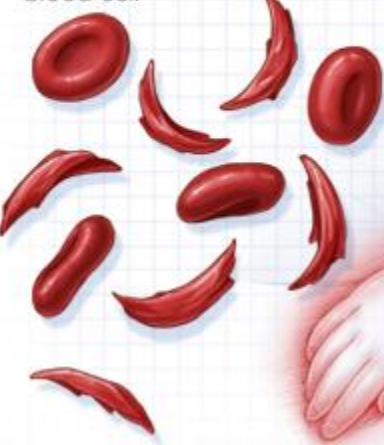


## Sickle Cell Anemia

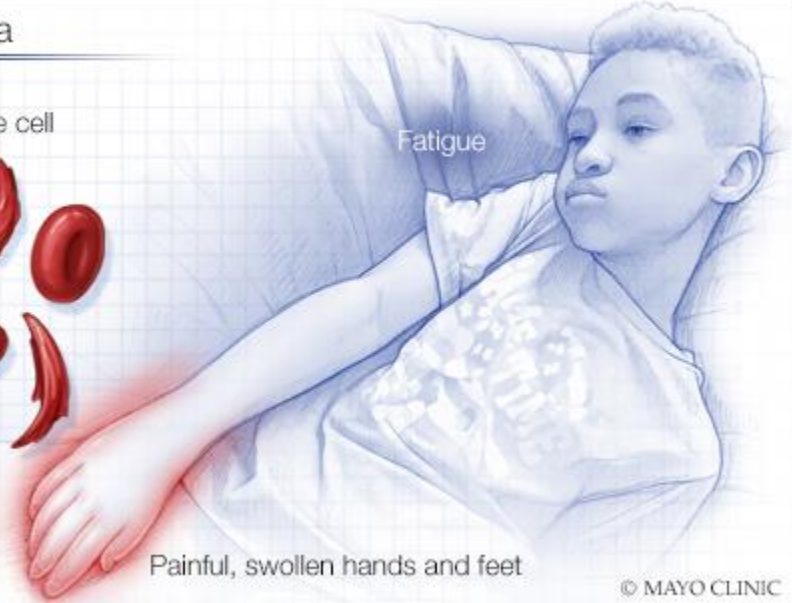
**Normal blood cell**



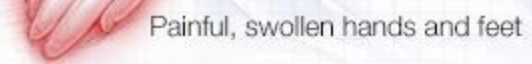
**Sickle cell**



**Fatigue**



**Painful, swollen hands and feet**



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## Symptoms of sickle cell anemia

### Accelerated

It's the closest planet to the Sun and the smallest

### Fatigue

Saturn is a gas giant and has several rings

### Distracted

Earth is the third planet from the Sun

### Dizziness

Venus is the second planet from the Sun

### Swollen hand

Neptune is the farthest from the Sun

### Irritable

Despite being red, Mars is a cold place



### Dactylitis:

- Painful swelling of hand and feet. Infarction of bone marrow due to occlusion of blood supply\*
- Manifests: 6 mos - 2 yrs
- Rx: Hydration, pain management

*\*painful swelling of the hands and/or feet as a manifestation of sickle cell disease in young children*