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Management of Sickle Cell Anaemia in Nigerian Public Hospital; the Nursing Perspective

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1. Introduction

Sickle Cell disease an inherited group of disorders that occur that occur as a result of misshaping of the red blood cells (RBC). Sickle Cell Anaemia on the other hand is inherited form of anaemia in which there are no enough RBC to carry adequate oxygen to all parts of the body.

For a baby to be born with sickle cell anemia, both parents must carry a sickle cell gene.

2. Definitions

Sickle Cell anemia is one of a group of disorders known as sickle cell disease. Sickle cell disease is an inherited red blood cell disorders in which there aren't enough healthy Red Blood Cells to carry oxygen throughout the body.

Normal red blood cells are rounded and disk-shaped. In sickle cell anemia, some red blood cells become deformed, so they look like sickles used to cut wheat. These unusually shaped cells give the disease its name.

3. Background

Sickle Cell Anaemia was said to be first discovered in 1904, Walter Clement Noel traveled from Grenada to the United States to start studying at the Chicago College of Dental Surgery. A few months later he was admitted to the Presbyterian Hospital in Chicago when he developed severe respiratory distress and a leg ulcer, both of which we now know are symptoms of sickle cell. Dr. Earnest E. Irons, the Intern who was on duty that day, performed a routine blood test and a urine analysis for Noel and was the first to observed these "pear shaped, elongated" sickled blood cells.

It was not until 1910 that Dr. James Herrick, the supervisor of Dr. Irons, published his article describing these "peculiar elongated and sickle shaped red blood corpuscles in a case of severe anemia." This was the first documented and recorded case of Sickle cell in Western medicine. Or Noel returned to Grenada in 1907 and ran his dental practice in St. Georges, the capital city, until he died at the age of 32 from the acute chest syndrome.

1917 - Genetic basis for SCD Dr. V. Emmel.

The third case of Sickle cell was described In 1915 by Cook and Meyer In a 21-year-old woman. Interestingly, blood samples from both the patient and her father, who displayed no symptoms, showed the sickling deformity of the red cells and three of her siblings had died from severe anemia.

These observations made by Dr. Emrnel suggested a genetic basis for the disease but also led to a period of confusion with the genetics of the disease.

1922 - Dr V.R Mason named the disease Sickle Cell Anemia. Dr Mason, who observed the fourth reported case of Sickle cell, was also the first to call the disease "sickle cell anemia" and to notice the similarities between the cases. He also noted that all of these patients were black, inadvertently giving rise to the popular misconception that sickle cell originated from people of African origin.

4. Incidence

As at 2015, according to WHO 4.4 million people in the world were said to have SCD. About 80% of cases are believed to occur in sub-Sahara Africa of which Nigeria is one. There are about 100,000 cases every year in Nigeria. Sickle cell anemia is most common in tropical Africans in people of African descent; about 1 in 10 African- American carries the abnormal gene. However, sickle cell anemia also appears in other ethnic populations, including people of Mediterranean or East Indian Ancestry. Overall, 1 in every 400 to 600 black children has sickle cell anemia.

5. Brief anatomy of the RBC

The normal RBC5 are round-shaped cells, highly flexible and contains an iron rich compound known as Haemoglobin (Hb) the compound which gives the blood its red colour. These cells move easily through the blood vessels to supply oxygen and nutrients to all the cell and tissues of the body.

The RBC is produced in the bone marrow and has a life span of about 120 days after which it dies away and replaced by new one.

• In SCD, there is a genetic mutation which signals the body to produce abnormal Hb, (valine glutamic Acid substitution). This abnormal Hb causes the RBCto become rigid, sticky and crescent moon shape otherwise called sickle shaped.

6. Causes

Sickle cell anemia is caused by a mutation in the gene that tells the body to make the iron-rich compound that makes blood red and enables red blood cells to carry oxygen from the lungs throughout the body (hemoglobin). In sickle cell anemia, the abnormal hemoglobin causes red blood cells to become rigid, sticky and misshapen. Both mother and father must pass the defective form of the gene for a child to be affected.

If only one parent passes the sickle cell gene to the child, that child will have the sickle cell trait. With one normal hemoglobin gene and one defective form of the gene, people with the sickle cell trait make both normal hemoglobin and sickle cell hemoglobin.

Their blood might contain some sickle cells, but they generally don't have symptoms. They're carriers of the disease, however, which means they can pass the gene to their children.

7. Predisposing factors

Cold temperature; Cold can aggravate the sickling process, because vasoconstriction slows the blood flow.

Tissue hypoxia; Tissue hypoxia and necrosis causes a type of sickle cell crisis called the sickle crisis.

Human parvovirusBl9 exacerbates aneamia by temporally suppressing bone marrow erythropoietic activity leading to transient aplastic crisis.

Splenic Infarction; Sequestration crisis results when other organs pool the sickled cells, just like the spleen.

8. Pathophysiology

In SCD, there is production of abnormal cells Hb, which makes the RBc to loose it's elasticity becomes rigid, sticky and sickled shape, these irregular shaped cells get stuck in small tiny vessels leading to vessel occlusion and ischaemia, blocking blood flow through the tiny capillaries which supply oxygen and nutrients to the cells and tissues. This brings about periodic episodes of pain, otherwise referred to as crisis, or vaso-occlusive crisis, especially to the chest, abdomen, joints, hands, feets and also in the bones.

9. Types

Hemoglobin SS disease

Hemoglobin SS disease is the most common type of sickle cell disease. It occurs when you inherit copies of the hemoglobin S gene from both parents. This forms hemoglobin known as HbSS. As the most severe form of SCD, individuals with this form also experience the worst symptoms at a higher rate.

Hemoglobin SC disease

Hemoglobin SC disease is the second most common type of sickle cell disease. It occurs when you inherit the Hb C gene from one parent and the Hb S gene from the other. Individuals with Hb SC have similar symptoms to individuals with Hb SS. However, the anemia is less severe.

Hemoglobin SB+ (beta) thalassemia

Hemoglobin SB+ (beta) thalassemia affects beta globin gene production. The size of the red blood cell is reduced because less beta protein is made. If inherited with the Hb S gene, the person will have hemoglobin S beta thalassemia. Symptoms are not as severe.
Hemoglobin SB 0 (Beta-zero) thalassemla:

Sickle beta-zero thalassemia is the fourth type of sickle cell disease. It also involves the beta globin gene. It has similar symptoms to Hb SS anemia. However, sometimes the symptoms of beta zero thalassemia are more severe. It is associated with a poorer prognosis.

10. Inheritance of sickle cell disease

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 gene and be completely unaffected.

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.

If one parent has sickle cell trait (HbAS) and the other has sickle cell anemia (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.

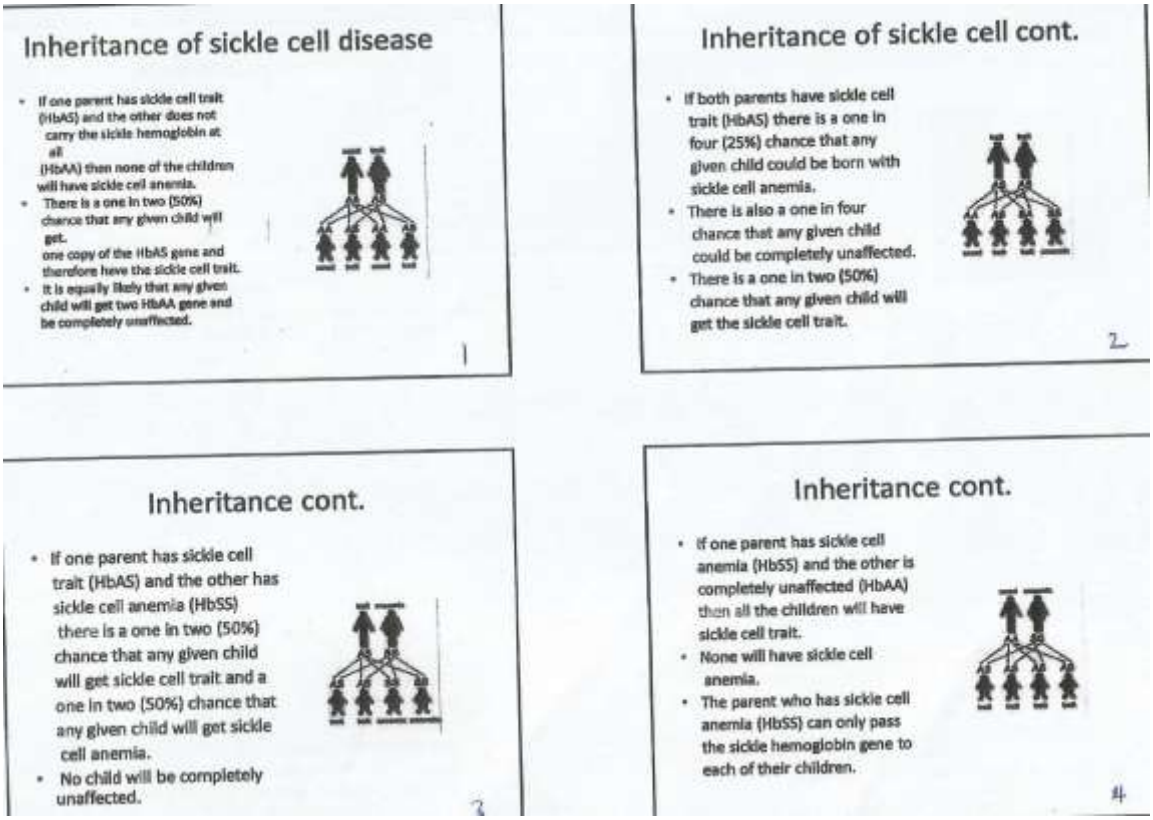
No child will be completely unaffected.

If one parent has sickle cell anemia (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.

Diagram below shows the four different cases



11. Signs and symptoms

Signs and symptoms of sickle cell anemia usually appear around 5 months of age. They vary from person to person and change over time.

Anemia Sickle cells break apart easily and die, leaving few red blood cells. Red blood cells usually live for about 120 days before they need to be replaced. But sickle cells usually die in 10 to 20 days, leaving a shortage of red blood cells (anemia).
Fatigue due to lack of oxygen

Episodes of pain: Periodic episodes of pain, called pain crises, are a major symptom of sickle cell anemia.

Pale skin or nail beds

Painful swelling of the hands and feet: The swelling is caused by sickle-shaped red blood cells blocking flow to the hands and feet.

Frequent infections: Sickle cells can damage an organ that fight infection (spleen), leaving the child more vulnerable to infection.

Fever: People with sickle cell anemia have an increased risk of serious infection, and fever can be the first sign of an infection.

Delayed growth or puberty: Red blood cells provide the body with the oxygen and nutrients needed for growth. A shortage of healthy red blood cells can slow growth in infants and children and delay puberty in teenagers.

Vision problems: Tiny blood vessels that supply the eyes can become plugged with sickle cells. This can damage the retina and can lead to vision problems.

ESR: Elevated.

Erythrocyte fragility: Decreased

Serum bilirubin (total and indirect): Elevated

Alkaline phosphatase: Elevated during vaso-occluslve crisis.

Serum potassium and uric acid: Elevated during vasoocclusive crisis (RBC hemolysis).

Serum iron: May be elevated or normal

Total iron-binding capacity (TIBC): Normal or decreased. Urine fecal urobilinogen: increased

12. Nursing management

For now there is no known cure for SCA but the condition can be managed.

The goals of management are, 1. Avoid crisis 2. To relieve symptoms.

1. To avoid crisis-

Avoid extreme weather condition (cold/stress.), Childhood vaccination against childhood diseases. Keep surroundings clean, to be free from mosquitoes, sleep under ITN (Insecticide treated net). Increase intake of fluids at least 2.5 liters of water per day, adequate diet rich in fruit and vegetables.

Routine iron supplement to boost production of RBC e.g. Folic acid (FA)

Intermittent preventive treatment for malaria — e.g. paludrine

Avoid over the counter drugs

Avoid too much stress.

2. To relieve symptoms

Nursing management of a child on admission is that of supportive care both for the child and care giver. Establish good rapport with the child and caregiver. Encourage them to ask questions. Educate the patient on disease process and provide clarification in clear terms, monitor record vital signs and put child in position most appropriate to reduce pain in the affected area. Also, provide gentle massage with warm compress on affected area to improve blood circulation if child is running temperature expose and tepid sponge. Encourage liberal fluid intake, Give more of fruits and vegetables, Maintain intake output chart, Monitor blood transfusion and reaction and report appropriately, Encourage passive exercises as child can tolerate, give prescribed drugs e.g. analgesics and antibiotics, On discharge encourage caregiver on follow up visits.

13. Genetic counseling

Chorionic villus sampling, or CVS, Involves the removal and testing of a very small sample of the placenta during early pregnancy. The sample, which contains the same DNA as the fetus, is removed by catheter or a fine needle Inserted through the cervix or by a fine needle Inserted through the abdomen. The tissue is tested for genetic changes identified in an affected family member. Results are usually available within 2 weeks.

Case study of a child with sickle cell anaemia

• Name: A.M

• Sex:female

• Age: 6yrs

• Date of adm: 25/1/2021

• Date of disc: 5/2/2021

• A six year female child with the above statistics reported into Emergency Paediatric Unit (EPU) with the history of fever, pains of lower and upper long bone, loss of appetite, general body weakness, stunted growth, skin eruptions and pale conjunctiva, with a pcv of 13%. Was seen by the Dr and a diagnosis of vaso-occlusive crisis? malaria in a known sickle cell disease patient.

The following nursing diagnosis were identified:

Nursing diagnosis

1. Acute pain.

2. Hyperthermia.

3. Impaired gaseous exchange.

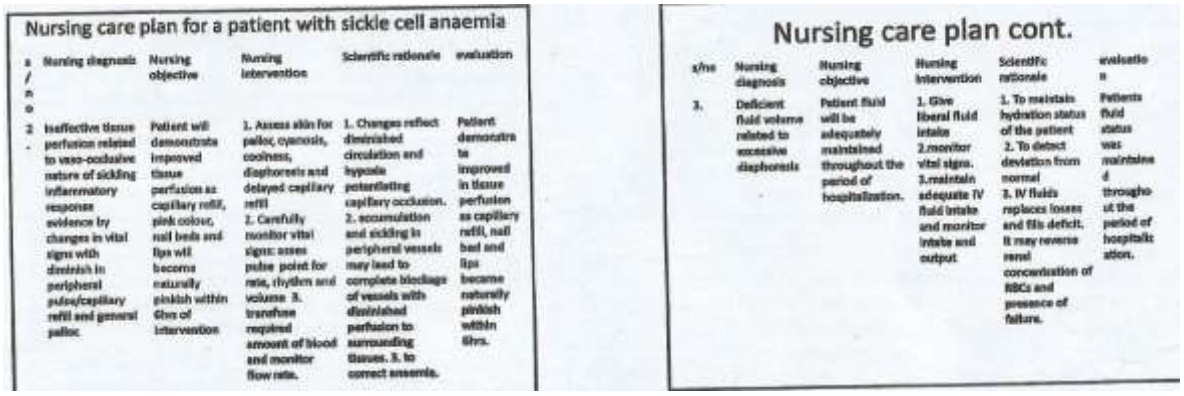
4. Ineffective tissue perfusion.

5. Risk for deficient fluid volume.

6. Risk for impaired skin integrity.

7. Deficient knowledge.

Nursing care plan of a child with sickle cell anaemia



Nursing care plan for a patient with sickle cell anaemia

s/no	Nursing diagnosis	Nursing objective	Nursing intervention	Scientific rationale	evaluation
2.	Ineffective tissue perfusion related to vaso-occlusive nature of sickling inflammatory response evidence by changes in vital signs with diminish in peripheral pulse/capillary refill and general pallor.	Patient will demonstrate improved tissue perfusion as capillary refill, pink colour, nail beds and lips will become naturally pinkish within 6hrs of intervention	1. Assess skin for pallor, cyanosis, coolness, diaphoresis and delayed capillary refill 2. Carefully monitor vital signs: asses pulse point for rate, rhythm and volume 3. transfuse required amount of blood and monitor flow rate.	1. Changes reflect diminished circulation and hypoxia potentiating capillary occlusion. 2. accumulation and sickling in peripheral vessels may lead to complete blockage of vessels with diminished perfusion to surrounding tissues. 3. to correct anaemia.	Patient demonstrate improved in tissue perfusion as capillary refill, nail bed and lips became naturally pinkish within 6hrs.

Nursing care plan cont.

s/no	Nursing diagnosis	Nursing objective	Nursing intervention	Scientific rationale	evaluation
3.	Deficient fluid volume related to excessive diaphoresis	Patient fluid will be adequately maintained throughout the period of hospitalization.	1. Give liberal fluid intake 2.monitor vital signs. 3.maintain adequate IV fluid intake and monitor intake and output	1. To maintain hydration status of the patient 2. To detect deviation from normal 3. IV fluids replaces losses and fills deficit. It may reverse renal concentration of RBCs and presence of failure.	Patients fluid status was maintained throughout the period of hospitalization.

14. Prevention

Genetic counseling for people to know their status of genotype before marriage, Health education in hospitals and clinic will do some help. Investigation i.e. sickling test should be done to know the genotype. Early therapy of folic acid therapy and penicillin administration can be the sure way.

15. Complications

Stroke: Sickle Cells can block blood flow to an area of the brain. Signs of stroke include seizures, weakness or numbness of the arm and legs, sudden speech difficulties, and loss of consciousness. If a child has any of these signs and symptoms, seek medical treatment immediately. A stroke can be fatal.

Acute chest syndrome: A lung Infection or sickle cells blocking blood vessels in the lungs can cause this life-threatening complication, resulting in chest pain, fever and difficulty in breathing. It might require emergency medical treatment.

Pulmonary hypertension: People with sickle cell anemia can develop high blood pressure In their lungs. This complication usually affects adults.

Shortness of breath and fatigue are common symptoms of this condition, which can be fatal.

Organ damage: Sickle cells that block blood flow to organs deprive the affected organs of blood and oxygen. In sickle cell anemia, blood is also chronically low in oxygen. This lack of oxygen-rich blood can damage nerves and organs, including the kidneys, liver and spleen, and can be fatal.

16. Medical Management

Blindness: Sickle cells can block tiny blood vessels that supply the eyes. Overtime, this can damage the eye and lead to blindness.

Leg ulcers: Sickle cell anemia can cause open sores on the legs.

Gallstones: The breakdown of red blood cells produces a substance called bilirubin. A high level of bilirubin in the body can lead to gallstones.

Priapism: In this condition, men with sickle cell anemia can have painful, long-lasting erections. Sickle cells can block the blood vessels in the penis, which can lead to impotence over time.

Pregnancy complications: Sickle cell anemia can Increase the risk of high blood pressure and blood clots during pregnancy. It can also Increase the risk of miscarriage, premature bIrth and having low birth weight babies.

17. Medical management

Folic Acid therapy daily may be initiated from birth to 5yrs, is recommended due to their immature immune system which makes them more prone to childhood illnesses.

Antibiotics — e.g. penicillin are usually given at every clinic visit.

Pain relieving medications like Paracetamol (pcm), pentazocin, ibuprofen, diclofenac and opiomare given.

Broad spectrum antibiotics are given. E.g. ceftriaxone when patient is on admission.

Blood transfusion with RBC only to increase the number of healthy RBC in circulation.

Exchange blood transfusion may also be carried out, involves exchange of significant portion of the child's RBC mass for normal RBC, which decreases the percentages of HbS.

Intravenous Fluid (IVF) therapy especially with normal saline to dilute the already thick, sticky blood in circulation.

Bone marrow transplant- offers the only potential cure for SCA. It is reserved for younger children of <l6yrs. The risk associated with the procedure increases with age, including death. To get a compatible donor is also difficult.

18. Genetic counseling

Two tests can be used to help expectant parents find out if their child is affected.

1. Amniocentesis, done usually at 14-16 weeks of pregnancy, tests a sample cf the amniotic fluid in the womb for genetic defects (the fluid and the fetus have the same DNA). Under local anesthesia, a thin needle is inserted through the woman's abdomen and into the uterus. About 20 milliliters of fluid (roughly 4 teaspoons) is withdrawn and sent to a lab for evaluation. Test results often take 1-2 weeks.

19. Prognosis

The average life expectancy in developed world is 40-6Oyears so in Nigeria being a developed country the life expectancy is less than 4Oyrs therefore the prognosis is not very encouraging.

20. Summary

Sickle cell anemia is a severe hemolytic anemia that results from the inheritance of the sickle hemoglobin gene. This gene causes the hemoglobin molecule to be defective. The sickle hemoglobin (HbS) acquires a crystal-like formation when exposed to low oxygen tension. The oxygen level in venous blood can be low enough to cause this change; consequently, the erythrocyte containing HbS loses its round, pliable, biconcave disk shape and becomes deformed, rigid, and sickle shaped. These adhere to the endothelium of small vessels; leading to reduced blood flow to a region or an organ. If ischemia or infarction results, the patient may have pain, swelling, fever, and sickle cell aneamia.

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