# Nursing Care of Children with Altered Hematological Function

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# Outline

1. Introduction
2. Anemia
3. Iron deficiency anemia
4. G6PD deficiency
5. Sickle cell anemia

# ANEMIAS--Generalities

- Definition:Condition in which the concentration of hemoglobin or the number of red blood cells are reduced below normal
- Anemia is not a specific entity but an indication of underlying pathologic processor disease,may normal size & color
- Rate of RBC production below that of cell destruction ,below the number of child age .

# ANEMIAS--Classification

- On an etiologic basis anemia result from:

1)Inadequate production of HB or RBC's which may be due to

a) lack in the BM of some substances necessary in the formation of cells( Iron,folic acid ….etc)

b) Decreased number of red cell precursors in the BM, which may be congenital or acquired

(eg. Toxic or Chemical agents)

2)Excessive loss of RBC's( hemolysis or hemorrhage)

# ANEMIAS-Tests in diagnosis 1

1) HB level and RBC's count

2) Hematocrite or packed RBC's volume ( the ratio of the volume of RBC's to the volume of whole blood)

# ANEMIAS- Tests in diagnosis 2

3) Red blood cells indices:

Mean corpuscular volume(MCV)=

(Hte/ RBC's count= normal 75-100 fimtoliter)

If >100 → macrocytic anemia

If < 75 → microcytic anemia

If 75-100 → normocytic anemia

# ANEMIAS Tests in diagnosis 3

Mean Corpuscular Hemoglobin( MCH) =

Average quantity of HB per individual red cell (HB/RBC's count =

Normal 27-31picogramme

# ANEMIAS-Tests in diagnosis 4

Mean Corpuscular Hemoglobin Concentration (MCHC)= Average concentration of HB in a volume of packet RBC's =

HB/Hematocrite = Normal 32-35 %

Both MCH and MCHC are used to determined the content of HB in RBC's

If normal MCH and MCHC = Normocytic anemia

If low MCH and MCHC =Hypochromic anemia

# ANEMIAS- Tests in diagnosis 5

4) Peripheral blood smear (film )

5) Reticulocytes count = reflect the state of activity of the BM (Normal value is 0.5-1.5% of the red blood cells )

Level < 0.5 represent inactive BM , High level represent BM regeneration

6) Other specific tests include = BM examination, HB electrophoresis, Osmotic fragility,Serum iron,B12,Folic acid…etc

# Normochromic

- Normocytic Anemia (normal color & size but have few number)

1. Blood loss
2. Anemia of acute infection
3. Anemia of renal disease
4. Anemia of Neoplastic disease

( malignant growth )

# A PLASTIC ANEMIA

- Inherited as an autosomal recessive trait (child with congenital abnormalities )
- Acquired a plastic anemia (radiations ,drug ,chemical ,chemotherapy )
- Assessment ( fatigue,anorexia,petechia ,GI bleeding)
- **HYPOPLASTIC ANEMIA**

( RBC only affect ) also genetic or acquired

# IRON DEFICIENCY ANEMIA

Etiology: Lack of iron in the diet or the child's inability to use the iron he ingest

1- Low birth weight, Prematurity, twins

=> decreased storage of iron

2- Continued or excessive milk administration without iron enriched food

3- Blood loss = Occult bleeding may be due to a lesion of the GI tract.

4- Malabsorption = ex: celiac disease

# IRON DEFICIENCY ANEMIA

## Clinical manifestations

- Pallor, irritability, anorexia, PICA( Ice,paper)
- Low HB, RBC’s count, & Low hematocrite
- Microcytic, hypochromic RBC’s ( reduction in the diameter of cell ,small size and pal color)
- Low serum iron, low saturation

# Clinical manifestations

- Enlarge heart
- Enlarge spleen
- Poor muscle tone
- Low level of activity
- Systolic murmur
- Decreased serum ferritin level
- finger spoon shape

# IRON DEFICIENCY ANEMIA

* TREATMENT

- Oral administration of simple ferrous salt:
  6 - 10 mg / kg / day of elemental iron
- Parenteral iron is seldom indicated
- Severely anemic children with HB level
< 4 gm / dl may be given blood transfusion
- Treat the underlining cause

# IRON DEFICIENCY ANEMIA

* PREVENTION

- Adequate diet include vegetables, meat and vitamins for full term infants
- Administration of iron (2 mg/kg) from the age of 2 months for low birth weigh and premature infants

# NURSING DIAGNOSIS

1. IMBALANCE NUTRITION less than body requirement r/t inadequate iron .

2. KNOWLEDGE DEFICIENT r/t cause and treatment of iron deficiency anemia.

# MEGALOBLASTIC ANEMIA
## Folic acid deficiency

* Etiology : 1. Decrease intake: Goat milk, excessive cooking

2. Decrease absorption: chronic diarrhea, phynotein

3. Increase demand: chronic hemolytic anemia

## Clinical manifestations

Macrocytic anemia (MCV >100), low reticulocyte count, large neutrophil with hyper segmented nucleus

## Diagnosis

Decreased serum folic acid,deceased red cell folate

## Treatment

Folic acid 2-5 mg /day for 3-4 weeks

# MEGALOBLASTIC ANEMIA
## Vitamin B 12 deficiency

Etiology

- Dietary deficiency is rare (Vegetarians)
- Failure to absorb vit B 12

Pernicious anemia

( absence gastric intrinsic factor)

Imerslund disease(defect absorption of vit B 12 )

## Clinical manifestations

Same as folic acid deficiency

## Diagnosis

Low serum vit B 12

## Treatment

Administration of vit B 12 (1-5 ηg/kg/day) for 2 weeks then once monthly 1 mg IM

# HEMOLYTIC ANEMIA

* The fundamental basis of the hemolytic anemia is a shortened survival time of the RBC,s(RBC normally spend 100-120 days in the circulation)
* Increased destruction of red cells result in anemia with increased quantity of heme pigment which may produces increased plasma concentration of HB (Hemoglobinemia/- uria) or increased conjugated serum bilirubine after catabolism of HB
* The normal BM response to hemolysis is by an increase of reticulocytes to > 2%

# SPHEROCYTOSIS 1

* Transmitted as an autosomal dominant trait
* The fundamental defect is an abnormality of the red cells membrane which permits excessive sodium ion entry into the cell

# SPHEROCYTOSIS 2

* Clinically,there is anemia, jaundice, splenomegaly,and presence of spherocytes in the blood film
* Diagnosis by increased osmotic fragility test
* Treatment by splenectomy (Preferably after the age of 4-5 years )

# G 6 P D deficiency 1

Glucose-6-phosphate Dehydrogenose

(Enzyme to maintenance of RBC life)

- Transmitted as a sex-linked recessive
- Mediterranean and middle eastern groups have high frequencies of G6OD deficiency

## Fig. 15.4 -- Punnett Square Analysis for G6PD Deficiency I

| | | **Mother's alleles** | |
|---|---|---|---|
| | | *Xn (normal)* | *Xn (normal)* |
| **Father's alleles** | *Xg (disease)* | *XgXn (female carrier)* | *XgXn (female carrier)* |
| | *Y* | *XnY (normal male)* | *XnY (normal male)* |

## Fig. 15.5 -- Punnett Square Analysis for G6PD Deficiency (II)

| | | **Mother's alleles** | |
|---|---|---|---|
| | | *Xn (normal)* | *Xg (disease)* |
| **Father's alleles** | *Xg (disease)* | *XgXn (female carrier)* | *XgXg (female with disease)* |
| | *Y* | *XnY (normal male)* | *XgY (male with disease)* |

# G 6 P D deficiency 2

## Clinically

Usually no evidence of hemolysis is apparent until 48-96 hours after the patient has ingested a substance which has oxidant properties

(Antipyretic,Sulfonamide, Anti malarial, or fava beans) producing an acute and severe hemolytic syndrome called FAVISM, Hb level become very low, presence of hemoglobinemia/-uria, mild jaundice, splenomegally and increased reticulocyte count

# G 6 P D deficiency 3

G6PD deficiency is an important cause of neonatal hemolysis and neonatal jaundice

# G 6 P D deficiency

## Diagnosis

Low G6PD activity in red blood cells

## Treatment

When hemolysis has occurred => Red blood cell transfusion

## Prevention

Avoiding ingestion of fava beans or oxidant substances

# Sickle cell anemia 1

- utosomal recessive abnormal shape (enlarge of RBC

Hb SA = normal

Hb SS = Abnormal Hb with valine substituted for glutamic acid in number 6 position of β chain

Sickling process is often initiated by low oxygen tension or low pH→ the sickle cell ????? Through capillaries→Vaso-occlusive crisis

# Sickle cell anemia 2

Clinically

= Moderate anemia

= Frequent articular and abdominal painful crisis

= Normocytic, normochromic anemia

= Peripheral red blood cells abnormality (sickle cell)

= Swelling of hand & feet

# Sickle cell anemia 2

= Hepatosplenomegaly
= Fever &back pain
= Sclera yellowed –decrease vision
= Anorexia

# Sickle cell anemia 3

## Diagnosis

Hb electrophoresis→Presence of Hb S

# Sickle cell anemia 4

Treatment

Instituted primarily for the crisis, the use of oxygen, the maintenance of good hydration, correction of acidosis, rest and sedative

Blood transfusion

BMT (stem cell transplantation)

# Nursing Diagnosis

- Ineffective tissue perfusion R/T generalize sickling

- Ineffective health maintenance R/T lack of knowledge

# Nursing Intervention

- Stresses management and family support
- HB level & blood transfusion
- Iron formula
- Immunization
- Regular school attend

# Aplastic and hypoplastic anemia

Pure red cell anemia

- Usually manifest in the first 4 months of life
- Severe normocytic, normochromic anemia,
- Very low reticulocyte, Normal WBC's and platelets count

Treatment…corticosteroid

# Fanconi anemia

- Familial aplastic anemia with a number of congenital anomalies
- Hematological manifestations rarely manifested prior to age 1 year by thrombocytopenia followed by neutropenia and anemia
- The most common congenital defects are skeletal (hypoplasia or absence thumb), renal, & brown pigmentation of the skin

# Treatment

Testosterone or other androgens with or without prednisone

Blood transfusion and chelationtherapy

BM transplantation

# Acquired or secondary aplastic anemia

Aplastic anemia

may occur as a reaction to various chemicals and drugs like chloramphenicol, sulfonamides, benzene, acetone, DDT ….etc