

Pediatric Hematology

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Approach to a Child with Anemia

Objectives

- Review a systematic approach to the differential diagnosis to a child with anemia
- Anemia
 - Review key aspects of history, physical and lab evaluation and put a plan for initial management

Hemoglobin

- ☐ Reversibly binds and transports O₂ from lungs to tissues
- ☐ Four globin chains & iron
 - Adult Hb (Hb A) $\alpha_2\beta_2$
 - Fetal Hb (Hb F) $\alpha_2\gamma_2$
 - Hb A₂ $\alpha_2\delta_2$
- The various globin chains differ in both the number and sequence of amino acids, and their synthesis is directed by separate genes.
 - Two sets of genes for α chains are located on chromosome 16.
 - The β , γ , δ genes are located on chromosome 11.

Fetal hemoglobin (Hb F)

- After 8 weeks of gestation, Hb F is the predominant Hb.
- At 6 months of gestation it constitutes 90% of the total Hb.
- At birth Hb F averages 50-65% of total Hb.
- Its synthesis rapidly declines after birth and by 6-12 months of age < 2% is present in the blood.

Hb A

- At birth, it averages 35-50% of the total Hb and by 6-12 months of age, adult Hb pattern appears.

Hb A₂

- Minor adult Hb component
- A: A₂ ratio is 30-1
- Normal value; 2-3.4%

Case presentation

- A 15 - month old child is seen at the primary Health center for MMR vaccination. The parents state she has been looking pale over the last 3 months .

- What else do you want to know?

Other symptoms that reflect severity of anemia (according to age), e.g. activity, sleep pattern, easy fatigability for older children.

- Signs of blood loss
- Dietary or feeding history, history of pica
- Family history of anemia, in other family members, splenectomy, gall stones at an early age.

Anemia

Anemia is defined by a laboratory value below the lower limit of normal (< 2 SD) for healthy children in any 1 of the 3 red cell indices:

- ✓ Hemoglobin level (Hb) ,
- ✓ Hematocrit (HCT)
- ✓ Red cell count (RBC)

The Hb, HCT and RBC levels vary with age:

- high in neonates
- falling to a low level at 2-3 months of age (physiological anemia of infancy)
- then rise steadily till puberty.

Morphological diagnosis of anemia

Red cell appearance

I. Microcytic (low MCV for the child age)

Children with microcytic anemia and low or normal reticulocyte counts most often have defects in erythroid maturation or ineffective erythropoiesis.

- Iron deficiency is the most common cause. Thalassemia trait constitutes the primary differential diagnosis when iron deficiency is suspected.
- Chronic disease or inflammation (more often normocytic), lead poisoning should also be considered.

Microcytosis and elevated reticulocyte counts are associated with thalassemia syndromes.

Normal values in relation to age

AGE	Hb Mean (lower limit)	HCT% Mean/ (lower limit)	MCV Mean/ (lower limit)
Newborn	16.5 (13.5)	51 (42)	108 (96)
1-12 months	12.6 (11.1)	37 (33)	78 (72)
2-6 Years	12.5 (11.0)	38 (34)	79 (75)
6-12 Years	13.5 (11.5)	39 (35)	81 (77)
12-14			
Male	14.0 (12.5)	43 (40)	84 (78)
Female	14.0 (12.0)	41 (36)	85 (79)

II. Normocytic (normal MCV)

Normocytic anemia and low reticulocyte count characterize many conditions;

- Anemia of chronic disease/inflammation
- Anemia associated with renal failure
- Decreased or absent red cell production secondary to infection, drugs, or endocrinopathy usually results in a normocytic anemia
- Bone marrow infiltration by malignancy.
- Acute bleeding

In children with normocytic anemia and an appropriate (high) reticulocytes, the anemia is usually a consequence of :

- Bleeding
- Hypersplenism,
- Ongoing hemolysis; resulting from conditions that are extrinsic (usually acquired) or intrinsic (usually congenital) to the red cell.
- Abnormal RBC morphology (e.g., spherocytes, sickle forms.

III. Macrocytic (MCV > 100 fl)

- Megaloblastic anemia (mainly folate deficiency, vitamin B12 deficiency).
- Other macrocytic anemias with low or normal reticulocyte counts include acquired and congenital aplastic anemias and hypothyroidism.

Pathophysiological mechanism of anemia

I- Defect in the production

II- Defect in the red cells

III- Increased destruction / Blood loss

Case presentation- Lab findings Cont.,

CBC: Hb 8.4 g/dl, Hct 23.4%, WBC 6,300, Plt 430,000 ,

MCV 54 fl, Red cell Distribution Width 17.8% .

Reticulocyte count is 1.8%.

S. Ferritin 6.5 ng/ml

The blood film shows microcytosis, hypochromia, and anisocytosis.

What is the most likely diagnosis?

What is your differential diagnosis?

How would you treat him?

How would you assess the response to treatment given?

Further readings

- 1. Illustrated textbook of Pediatrics by Tom Lissauer and Graham Clayden**
- 2. Nelson Textbook of Pediatrics**
- 3. Forfar & Arneils Textbook of Pediatrics.**