Pediatric Hematology - 2

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Megaloblastic Anemia

Etiology

- Inadequate dietary intake
- Decreased folate absorption e.g. celiac disease
- Congenital abnormalities in folate metabolism
- Drug-induced abnormalities in folate metabolism

Clinical features

- **In infancy**

- Pallor, irritability, failure to gain weight, chronic diarrhea.
- Hemorrhage in advanced cases (thrombocytopenia)

- **In older children**

Pallor and other features of anemia.

Vitamin B 12 deficiency

- Glossitis, vomiting, and jaundice.
- Neurological symptoms; paresthesia, sensory deficits, hypotonia, seizures, developmental delay.

Laboratory findings

- Macrocytic anemia
- Low reticulocyte count
- WBC: neutropenia, hypersegmentation of PMN (> 5 lobes)

- Thrombocytopenia
- Folate l Vitamin B 12 assay
- S. LDH is elevated

Treatment depends on the cause

Hemolytic Anemias

Hemolysis

Premature destruction of red cells.

Classification

During hemolysis;

- RBC survival is shortened,
- RBC count falls,
- Erythropoietin is increased,
- Stimulation of marrow activity results in increased RBC production *(increased percentage of reticulocytes in the blood).*
- Excessive red cells destruction leads to increased unconjugated bilirubin & urine urobilinogen.
- Symptoms include pallor, jaundice & splenomegaly.
- Pigment gall stones may complicate the condition.
- Aplastic crises; precipitated by parvo virus or folate deficiency.
- Clinically there is prominent maxillary bones, bossing of skull & on X-ray: radial striations (hair-on-end appearance)

Glucose-6-phosphate dehydrogenase deficiency

Etiology

- Inherited as an X-linked recessive gene.

❑ Is responsible for 2 clinical syndromes,

➢ Episodic hemolytic anemia induced by infections, certain drugs or, fava beans.

➢ Spontaneous chronic non-spherocytic hemolytic anemia.

➢ Neonatal jaundice

❑ Incidence 1-35 % in Mediterranean races.

Agents Precipitating Hemolysis in G-6-PD Deficiency

Antibacterials	Antimalarials	Others	Illness
Sulfonamides	Primaquine	Phenacetin	Diabetic acidosis
Trimethoprim-sulfamethoxazole	Chloroquine	Vitamin K analogs	Hepatitis
Nalidixic acid	Quinacrine	Acetylsalicylic acid	Sepsis
Chloramphenicol		Benzene	Fava beans
Nitrofurantoin		Naphthalene	

Laboratory data

- Fall in Hb level.
- Increase in reticulocyte count.
- Red cells reveal Heinz bodies.
- Direct measurement, enzyme activity in affected persons is ≤10% of normal

Treatment

- Prevention of hemolysis.
- When hemolysis occur, blood transfusion.

* **Recovery is the rule.**

Sickle Cell Disease

- An inherited disease of red blood cells (Autosomal recessive), caused by a mutation in the β-globin gene in which the 17th nucleotide is changed from thymine to adenine and the sixth amino acid in the β-globin chain becomes valine instead of glutamic acid.
- Polymerization of hemoglobin leads to a cascade of effects decreasing blood flow.
- Tissue hypoxia causes acute and chronic damage.
- Sickle cell anemia (SCA) can be diagnosed antenataly using amniocyte or chorionic villus DNA analysis.

Sickle cell trait

- Is usually associated with a benign clinical course.
- The amount of Hb S in sickle cell trait is < 50%.
- RBCs resist invasion by malarial parasites.
- Diagnosis - Hb-electrophoresis.

Complications

- ✓ Sudden death during vigorous exercise ،
- ✓ Splenic infarcts at high altitude،
- ✓ Hematuria،
- ✓ Hyposthenuria ،
- ✓ Bacteriuria .

Sickle Cell Anemia (Homozygos Hb S)

Severe chronic hemolytic anemia, resulting from premature destruction of RBCs.

Clinical manifestations

- Anemia gradually develops over the first 2-4 months of life.
- Acute dactylitis **(hand-foot syndrome)** is usually the first evidence of the disease in infants.
- The clinical course is associated with intermittent episodic events **(crises).**
  - ✓ Acute painful episodes
  - ✓ Acute splenic sequestration
  - ✓ Aplastic crisis
- Neurologic complications
- Acute chest syndrome

- Renal function is impaired

- Autosplenectomy- spleen is rarely palpable after 6 years of age.

- Cardiomegaly

- Pulmonary hypertension

- Increased susceptibility to infections

- Gall stones.

- Zinc deficiency.

- Poor growth & delayed maturation.

- Priapism

- Leg ulcers