

Pediatric Hematology - 4

Professor Meaad K. Hassan
Department of Pediatrics
Basrah Medical College

Hereditary spherocytosis

Etiology

- ❖ Autosomal dominant condition.
- ❖ There is abnormality of spectrin or ankyrin (major components of membrane), that are responsible for RBC shape.

Clinical features

- ▶ Hemolytic disease of newborn.
- ▶ In infants & older children:- anemia, splenomegaly, gall stones.
- ▶ Aplastic crises.

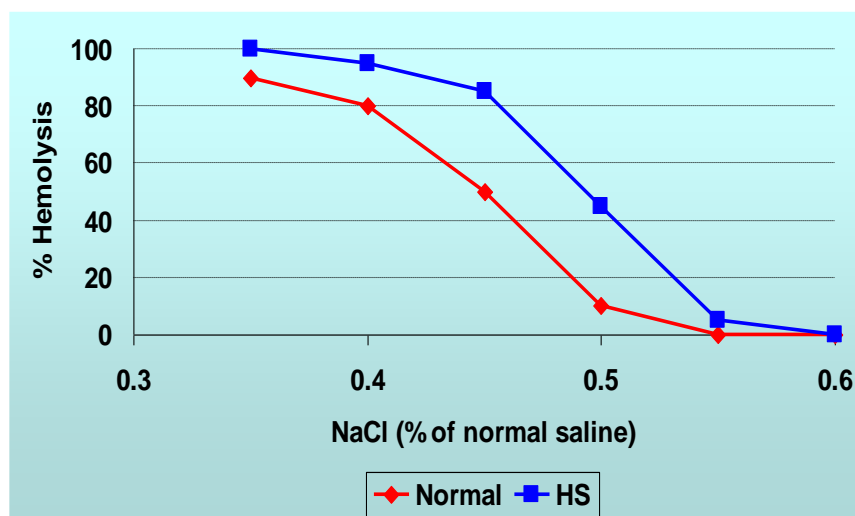
Lab. Findings

- ▶ Hb 6-10 gm/dl.
- ▶ Reticulocyte count 6-20%.
- ▶ Elevated s. unconjugated Bilirubin.

- ▶ MCV normal.
- ▶ MCHC (36-38 gm/dl red cells).
- ▶ Blood film:- **Spherocytes (15-20 % of red cells)**
- ▶ U/S: gall stone

Osmotic fragility test

- The RBCs are incubated in progressive dilutions of an iso-osmotic buffered salt solution.
- Exposure to hypotonic saline causes the RBCs to swell, and the spherocytes lyse more readily than biconcave cells in hypotonic solutions




If the diagnosis is less certain: flow cytometric EMA (eosin-5-maleimide) binding test and the cryohemolysis test.

Treatment

- ▶ Splenectomy (Laparoscopic splenectomy)
- ▶ Folic acid (1 mg/day)

Aplastic anemia

- ▶ Reduced red cell production
- ▶ In most types; WBC & platelets productions are also affected
 neutropenia & thrombocytopenia

Acquired

Constitutional

Acquired aplastic anemia

Etiology:-

RADIATION, DRUGS AND CHEMICALS

- ▶ Predictable: chemotherapy, benzene
- ▶ Idiosyncratic: chloramphenicol, antiepileptics, gold

VIRUSES

- Cytomegalovirus
- Epstein-Barr
- Hepatitis B
- Hepatitis C
- Human immunodeficiency (HIV)

PAROXYSMAL NOCTURNAL HEMOGLOBINURIA

IDIOPATHIC

Clinical features

- Increased risk of fatigue, infections, bleeding & heart failure.
- Spleen & Lymph nodes are not enlarged.

Differential diagnosis

- Collagen vascular diseases
- Malignancies
- Myelofibrosis
- Infections

Lab. Data

- ▶ Anemia
- ▶ Leucopenia (neutrophil C < 500/mm³).
- Platelet C. < 20,000/mm³
- ▶ Reticulocyte C < 1%
- ▶ **Bone marrow aspirate : & biopsy:**
 - Hypocellular

Treatment:-

➤ Supportive

A- Platelets transfusion

B- Antibiotics.

C- Blood transfusion

➤ Specific

- Allogeneic bone marrow transplantation (BMT) offers a 90% chance of long-term survival.
- Antithymocyte globulin (ATG) and cyclosporine (response rate of 60–80%)

References

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