Pediatric Hematology - 3

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Sickle Cell Anemia

Laboratory data

- Hb % : 5-9 gm / dl.
- Peripheral blood:- target cells, poikilocytes, irreversibly sickled cells.
- Reticulocyte count 5-15%.
- WBC 12,000-20,000/ cum.
- Abnormal liver function tests.
- Hb-electrophoresis :Hb S 80-95%, Hb F 2-20%

Treatment

- Prevention of complications
- Full immunization status
  - ✓ Polyvalent pneumococcal v .
  - ✓ H. influenzae v.
  - ✓ Hepatitis b v.
- Oral penicillin starting from 4 months -5 years of age.
- Parental education.

Immediate medical consultation if patient develops a temp. > 38.5C , severe pain or pallor.

<u>Painful episodes:</u>

- Oral acetaminophen (alone or with codeine).
- In severe episodes- Hospitalization, Anti-inflammatory agents.

- Correction of dehydration or acidosis.
- Narcotic analgesics

Blood transfusions in -:

- Disabling chronic pain.
- Ischemic organ damage.
- Stroke.
- Preparation for major surgery
- Sequestration crises.
- Aplastic crises.

Chemotherapy to stimulate Hb F synthesis ‹

Hydroxyurea

- Bone marrow transplantation

Thalassemia syndromes

- A heterogeneous group of inherited anemias characterized by defects in the synthesis of one or more of the globin chain subunits of the hemoglobin tetramer.
- *Classification*
  - ✓ α-Thalassemias
  - ✓ ß-Thalassemias
  - ✓ Delta Thalassemia

- Thalassemia is an autosomal recessive blood disorder.
- The primary defect is gene mutation that leads to inhibition of Hb synthesis and accumulation of intracellular iron.

ß-Thalassemias

β-thalassaemia trait

No symptoms

Mild anemia

Red cells are hypochromic microcytic with poikilocytosis.

- MCV is low (65 fl)
- MCH is low (< 26)
- S. Iron is normal or elevated

It is often misdiagnosed as iron deficiency anemia

Diagnosis

Hb-electrophoreses	**HbA2**	**3.4 -7%**
50% have slight increase in	**HbF**	**2- 6%**

β -Thalassemia Intermedia

- Globin chain production is moderately impaired.
- Has a wide spectrum of clinical phenotype.
- Patients maintain a satisfactory hemoglobin level of at least 6-7 g/dl at the time of diagnosis without the need for regular blood transfusions.

β-thalassaemia major

Severe progressive hemolytic anemia that becomes symptomatic in 2nd 6 months of life.

Clinical features

- ❑ Pallor
- ❑ Without regular blood transfusions, hypertrophy of erythropoietic tissues occur causing thin bones, pathologic fractures & characteristic facial appearance.

- Liver & spleen are enlarged.
- Impaired growth & delayed puberty.
- Diabetes mellitus.
- Cardiac complications

Laboratory data

- ✓ Hb falls < 5 gm/ dl unless blood is given.
- ✓ RBC: hypochromic, microcytic with fragmented poikilocytes & target cells**.**
- ✓ Elevated unconjugated bilirubin
- ✓ Hb F level > 70% of total Hb

Treatment

- ❖ Regular blood transfusions to maintain Hb level > 10 gm/ dl.
- Packed RBCs; 10-15 ml/kg over 2-3 hours.
- ❖ Treatment of iron overload by ***iron chelating agents***:-
- ❖ S. ferritin should be maintained ≤ 1000 mg/dl.
- Exjade® - deferasirox

Indications: patients ≥ 2 years, with chronic iron overload due to repeated blood transfusions . It is given orally, once daily.

- Deferoxamine (Desferal)

  Given through a pump S.C (30-50mg/kg) over 8-12 hr for 5-6 nights/ week. Vitamin C is given at the same time desferal injection.

- Deferiprone

- **Splenectomy**

Indications: - Size of the spleen

- Hypersplenism

- **Bone marrow transplantation**:

3 risk factors affect the outcome bone marrow transplantation:-

a- Hepatomegaly.

b- Portal fibrosis

c- Irregular iron chelation

References

1. Illustrated textbook of Pediatrics by Tom Lissauer and Graham Clayden

2. Forfar & Arneils Textbook of Pediatrics

3. Nelson Textbook of Pediatrics