TUTORIAL 14

EXAMINATION OF HAEMATOLOGICAL SYSTEM

OVERALL OBJECTIVES

At the end of this module, student should be able to

1) Recognise and manage the common haematological problems of neonates, infancy and childhood.
2) Interpret the FBC and state the changes that occur from birth to puberty
3) Recognise by appropriate history taking, clinical examination and relevant laboratory investigations, and approach to the following anaemias:
   - Iron deficiency
   - Folate deficiency
   - B12 deficiency
   - Anaemia of chronic disease
   - Anaemia infection
   - Aplastic anaemia
   - Haemolytic anaemias
4) Tabulate important aspects in prevention and management of above anaemias.
5) **Bleeding disorders / purpura**
   - State and recognise clinically the difference in the clinical presentation of vascular vs platelet vs co-agulation defects
   - Identify important aspects in the history (including patterns of inheritance) of a patient with a bleeding disorder
   - Tabulate important aspects in the aetiology, pathology and management of thrombocytopenia (ITP), haemophilia, A, B, DIC, Henoch Schonlein purpura and haemorrhagic disease of the newborn.
   - Interpret the basic screening tests and their application in the approach to a patient with a bleeding disorder.
6) **Malignancy**
   - Recognise the presenting features (clinical + laboratory) of leukaemia + lymphoma in childhood
   - State the differential diagnosis of above
   - State the steps in the management of these patients
   - Counsel the parents on effects of malignancy (including prognosis) and of cytotoxic therapy
7) **Component therapy**

   List indications for and complications of blood component therapy
8) **Ethics**
   - Malignancy
   - Cytotoxic use

CONSIDER HAEMATOLOGICAL SYSTEM INVOLVEMENT IF FOLLOWING HAS BEEN FOUND IN ABOVE EXAMINATION

- **Pallor:** long list of anaemias: microcytic, macrocytic, normocytic (low Retics, high Retics)
- **Petechiae:** purpura, ecchymosis: ALL, AML, aplastic anaemia, ITP
- **Pigmentation:** thalassaemia
- **Eczema:** Wiskott-Aldrich (with ear discharge)
- Chronic lung disease stigmata
- Joint swelling:
    - Haemophilia
    - Henoch-Schonlein purpura
    - Sickle cell anaemia, leukaemia
    - Irritable bowel disease
    - Juvenile idiopathic arthritis
- **Squint, nystagmus:** Fanconi's anaemia, 6th nerve palsy with IC bleed
- **Ptosis:** Fanconi's anaemia
- **Ataxic gait:** Vit B 12 def
- **Hemiplegia:** haemophilia with IC bleed, sickle cell anaemia with cerebral sickling
- Splenomegaly
    - Haemoglobinopathies
    - Malignancy
    - Infection – SBE
    - Osteopetrosis
    - Storage diseases
    - Congenital spherocytosis
- **Hepatomegaly:** as above, plus
    - Hepatitis
    - Wilson's disease (decrease in clotting factors 1, 2, 5)
- **Delay in Tanner staging:** Thalassaemia, sickle cell anaemia
- **Posterior iliac crest tenderness, scar:** bone marrow aspiration

THEN THINK SPECIFICALLY ABOUT SEX, RACE AND SYNDROMES

- Blackfan-Diamond red cell aplasia
- Fanconi's anaemia
- Thrombocytopenia absent radius
- Wiskott-Aldrich syndrome
- Dyskeratosis congenita
- Triade may become apparent in 1st 10 yrs of life (XLR 85%, AD & AR 15%)

- Reticulate lacy pigmentation of upper body
- Mucosal leukoplakia
- Nail dystrophy
- Swaschman-Diamond syndrome
- Pearson syndrome
    - Refractory sideroblastic anaemia
    - Cytoplasmic vacuolization bone marrow precursors
    - Metabolic acidosis
    - Mitochondrial DNA mutation
    - Exocrine pancreatic insufficiency