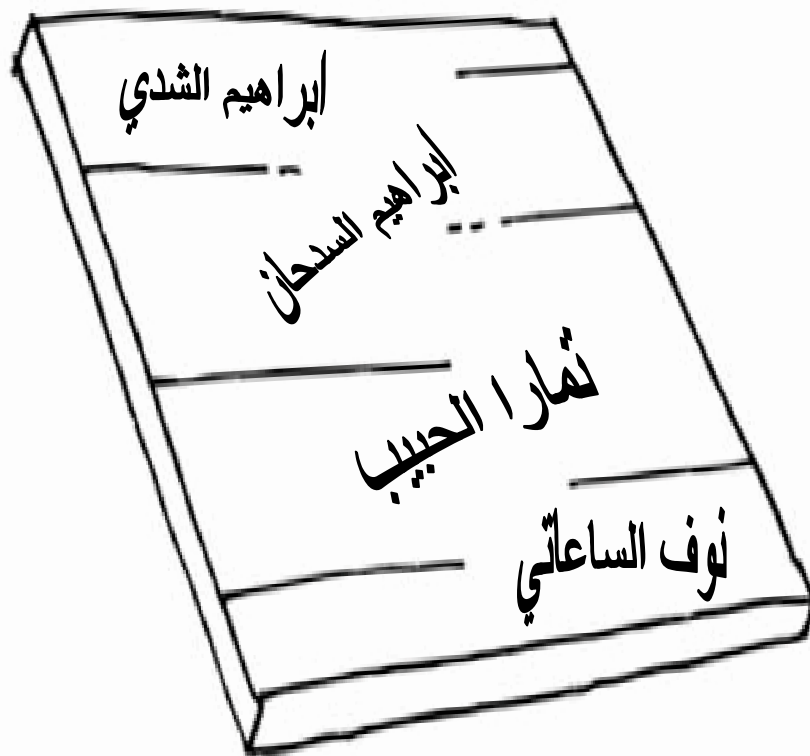
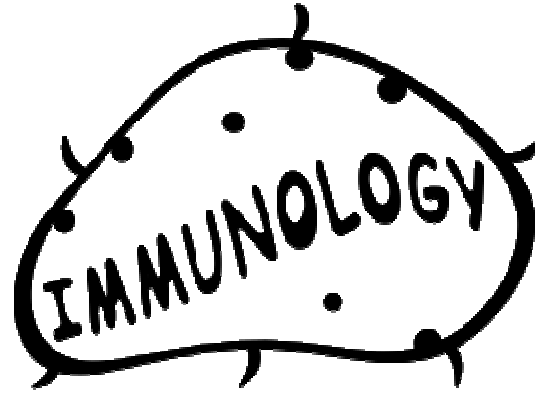




Immune Team ... !



# Immunodeficiency Diseases

- The most common cause of secondary immunodeficiency (acquired) in children is malnutrition (and it may occur in adults as well).
- Nephrotic syndrome causes immunodeficiency because patient losses proteins (mainly albumin and less  $\gamma$  globulin) in urine.
- Immunodeficiency diseases affects various components of the immune system such as lymphocytes (B or T) or even certain immunoglobulins or complement proteins.

## B cell diseases:

### *X-linked gammaglobulinaemia (Bruton's disease):*

- The most common B lymphocyte disorder
- Development of B cells 'maturation' is stopped because of a defect in an enzyme called Burton's Tryosine Kinase (BTK).
- \* BTK is an enzyme needed for the maturation of B cells to plasma cells.

### *Immunoglobulin disorders:*

IgA deficiency → could be a side effect of anti- convulsant drugs used to treat epilepsy (ex: Phenytoin).

Hyper-IgM syndrome → caused by a defect in the receptor (CD40L) on T helper cells.

## T cell diseases:

### *DiGeorge Syndrome:*

- Congenital absence of the Thymus gland → immature T cells → no CMI and HI is partially affected because of the absence of T helper cells.
- Accompanied by characteristic facial features = large forehead and low ears.
- Treatment is thymus transplantation with steps to prevent GVH.

GVH= Graft Vs Host = the graft produce lymphocytes that attack the host tissues.

## Severe combined immunodeficiency:

### *Deficiency of the enzymes ADA & PNP:*

- ADA & PNP are enzymes that degrade toxic metabolites of lymphocytes → absence of these enzyme leads to death of lymphocytes.

## Natural immunity disorders:

### *Chronic granulomatous disease:*

- Phagocytic cells kill microbes by substances that contain oxygen (nitric oxide, superoxide ,,,) → failure of phagocytic cells to take oxygen make them unable to make these substances "lose their weapons" → inability to kill microbes → granuloma formation.

### Complement deficiencies:

(C1, C2, C3, C4, C5, C6, C7, C8, C9) complement proteins There are

and C3 activates C2 and the C2 is activated by a stimulus it activates C1 When C1 ..... and so on C4 activates C3

⇒⇒ excessive complement production inhibitor enzyme will lead to deficiency of C (activation) and that shouldn't be happening in healthy people,