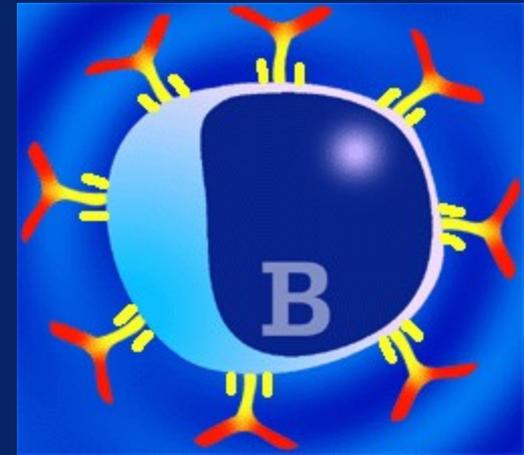


A collection of B cells, which are a type of white blood cell, shown in various sizes and orientations. The cells are rendered in a light blue, semi-transparent style against a dark blue background. Some cells are large and elongated, while others are smaller and more rounded. The text "B CELL IMMUNODEFICIENCY" is overlaid in the center in a bold, black, serif font.

**B CELL
IMMUNODEFICIENCY**

INTRODUCTION TO B CELL

- ✓ B lymphocytes, named after their site of origin in the bursa of Fabricius in birds or in the bone marrow in humans, form the basis for humoral immunity by their production of immunoglobulins.
- ✓ It belongs to adaptive immunity
- ✓ Antigen specificity
- ✓ Humoral immunity
- ✓ Final destination for B cell : Plasma cell



Cellular And Humoral Immunity

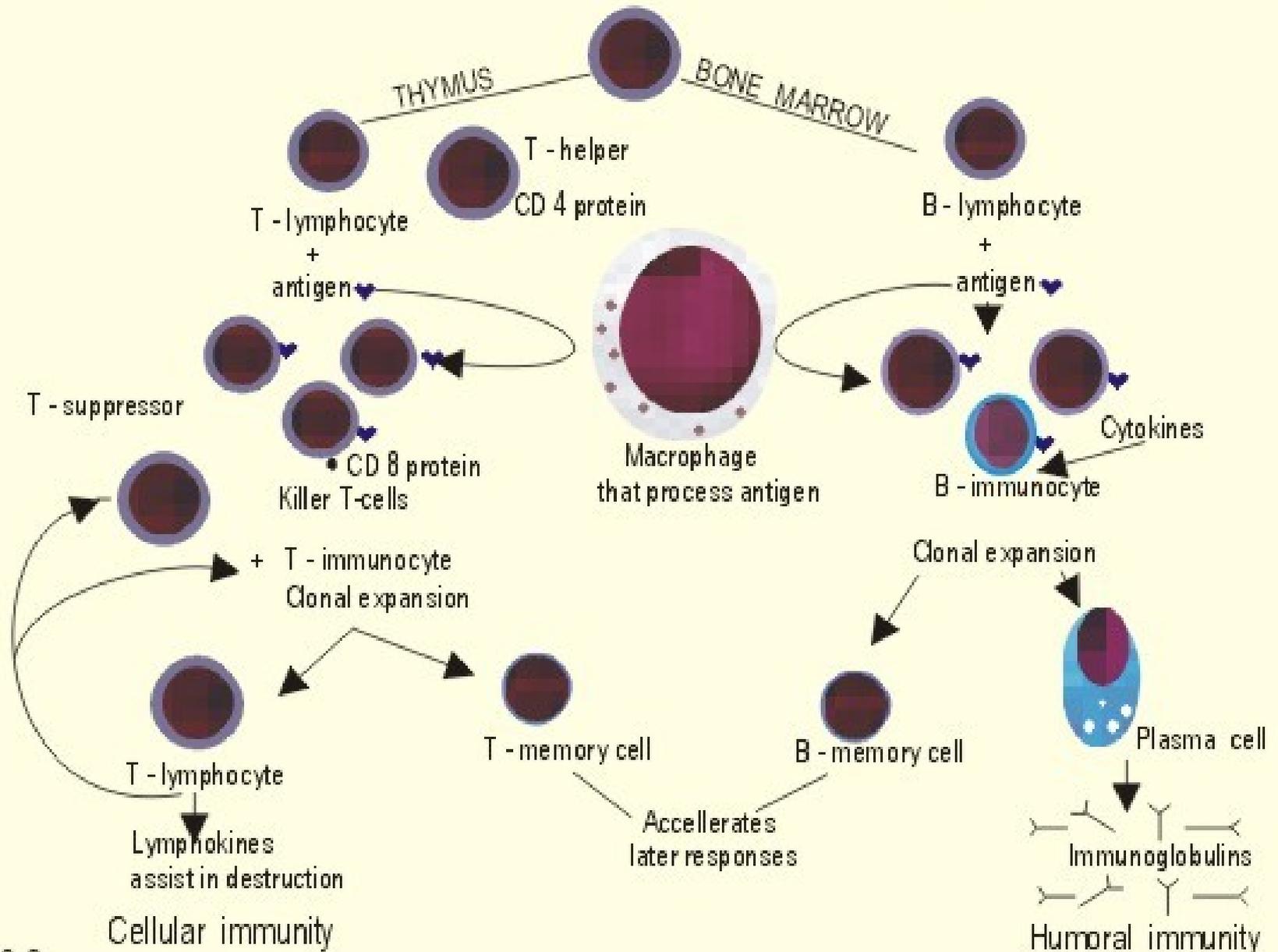
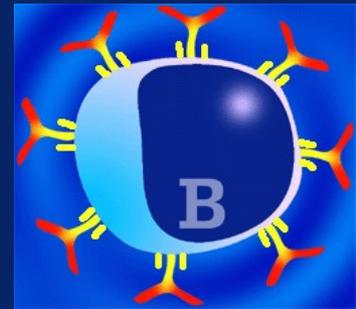


Fig. 32-3

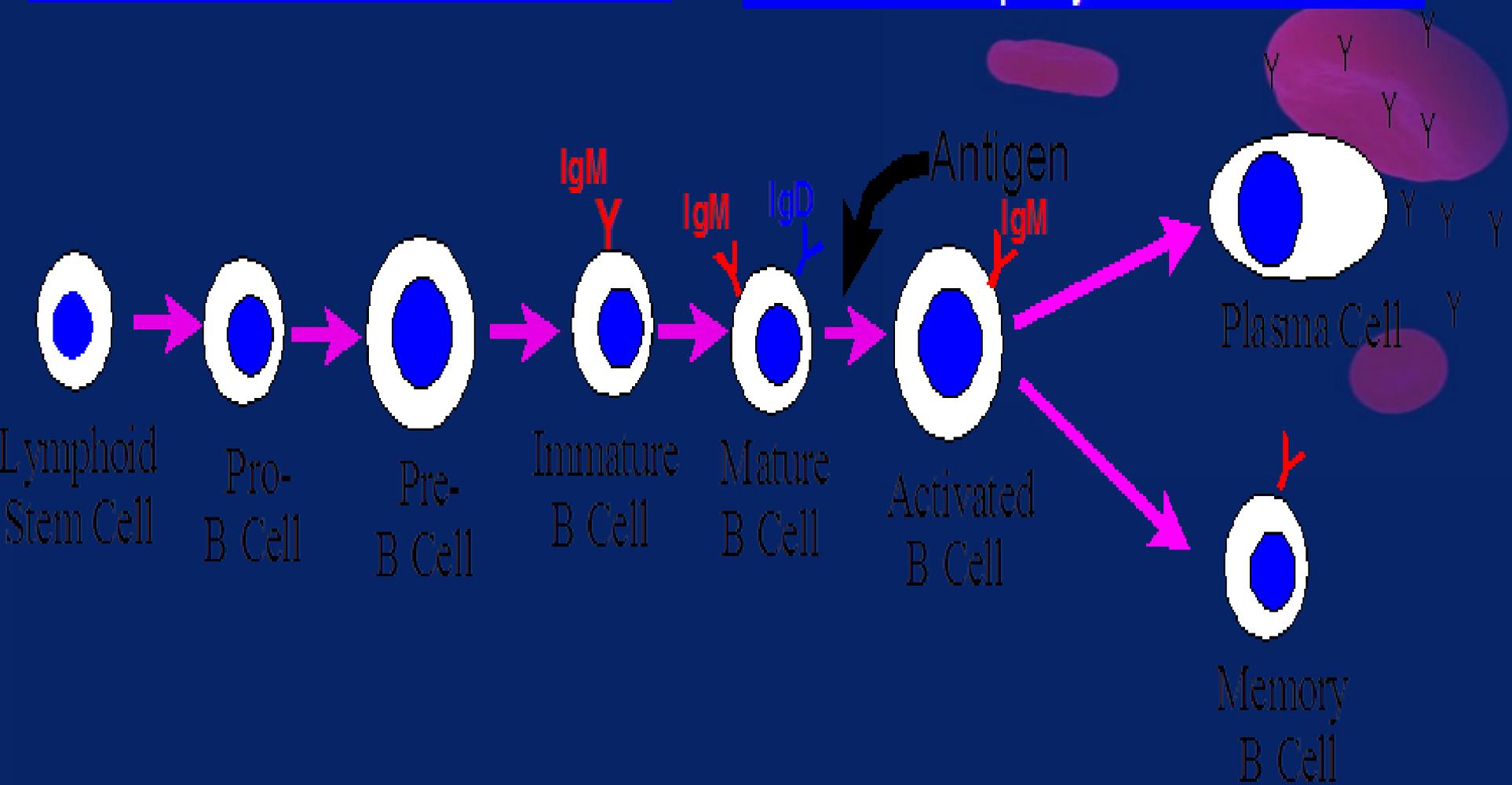
DEVELOPMENT OF B CELL

- ✓ The differentiation of B cells occurs in the *bone marrow* throughout the life of an individual.
- ✓ The earliest recognizable cell in the B cell lineage: pro B cell, followed by pre B cell
- ✓ Immature B cell: the cells which express IgM on its surface.
- ✓ Mature B cell: the cell which express IgM and IgD on its surface.



Bone Marrow

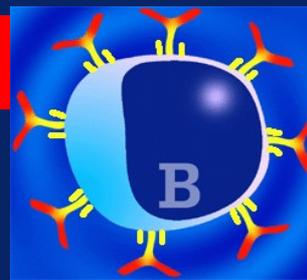
Periphery



Antigen Independent

Antigen Dependent

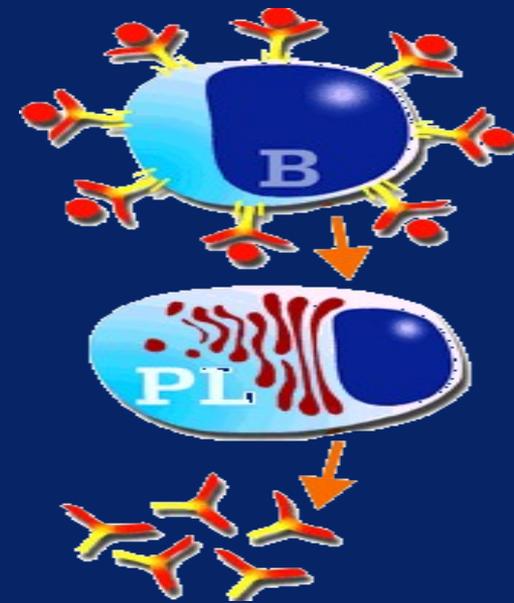
Primary Lymphoid Organ

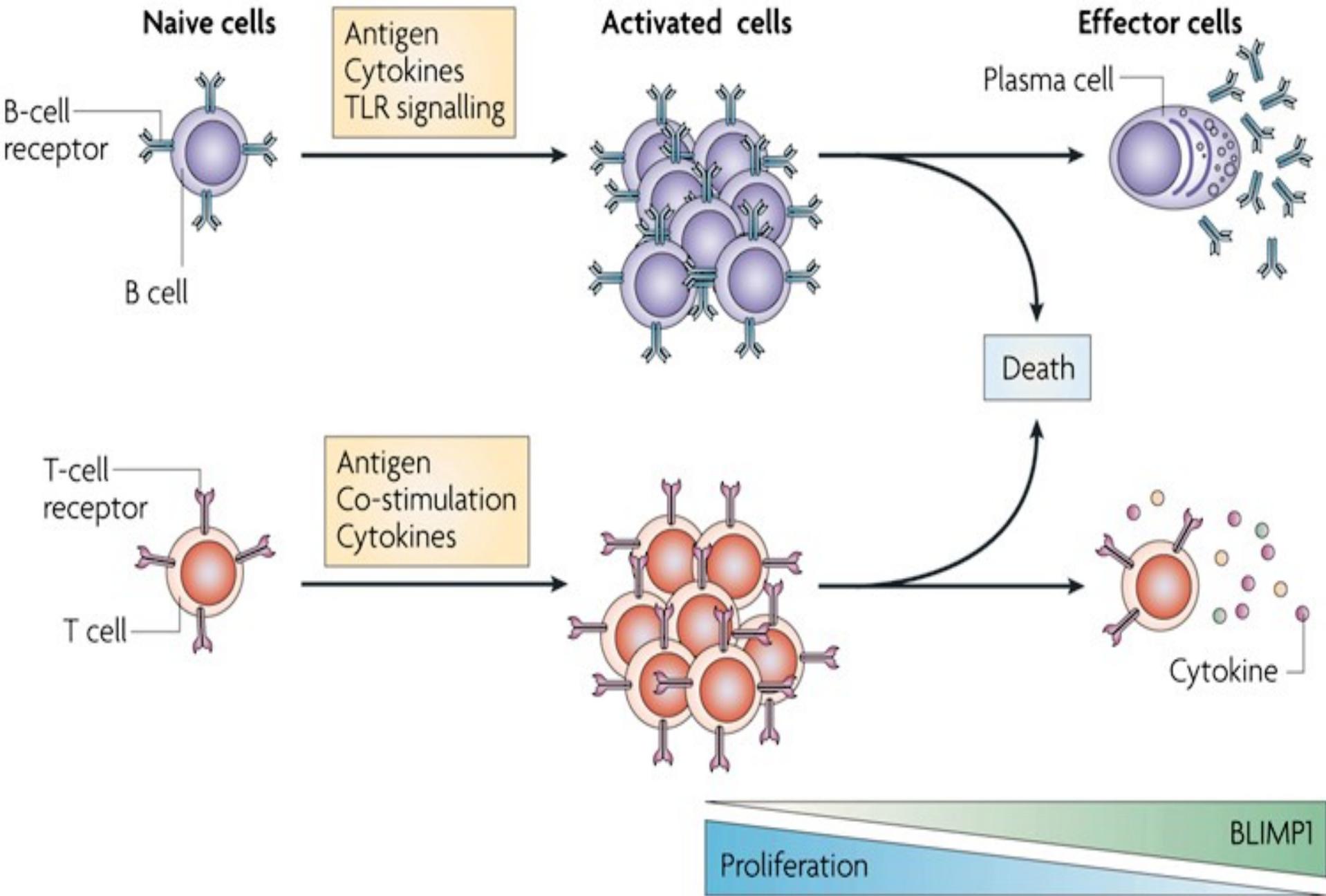


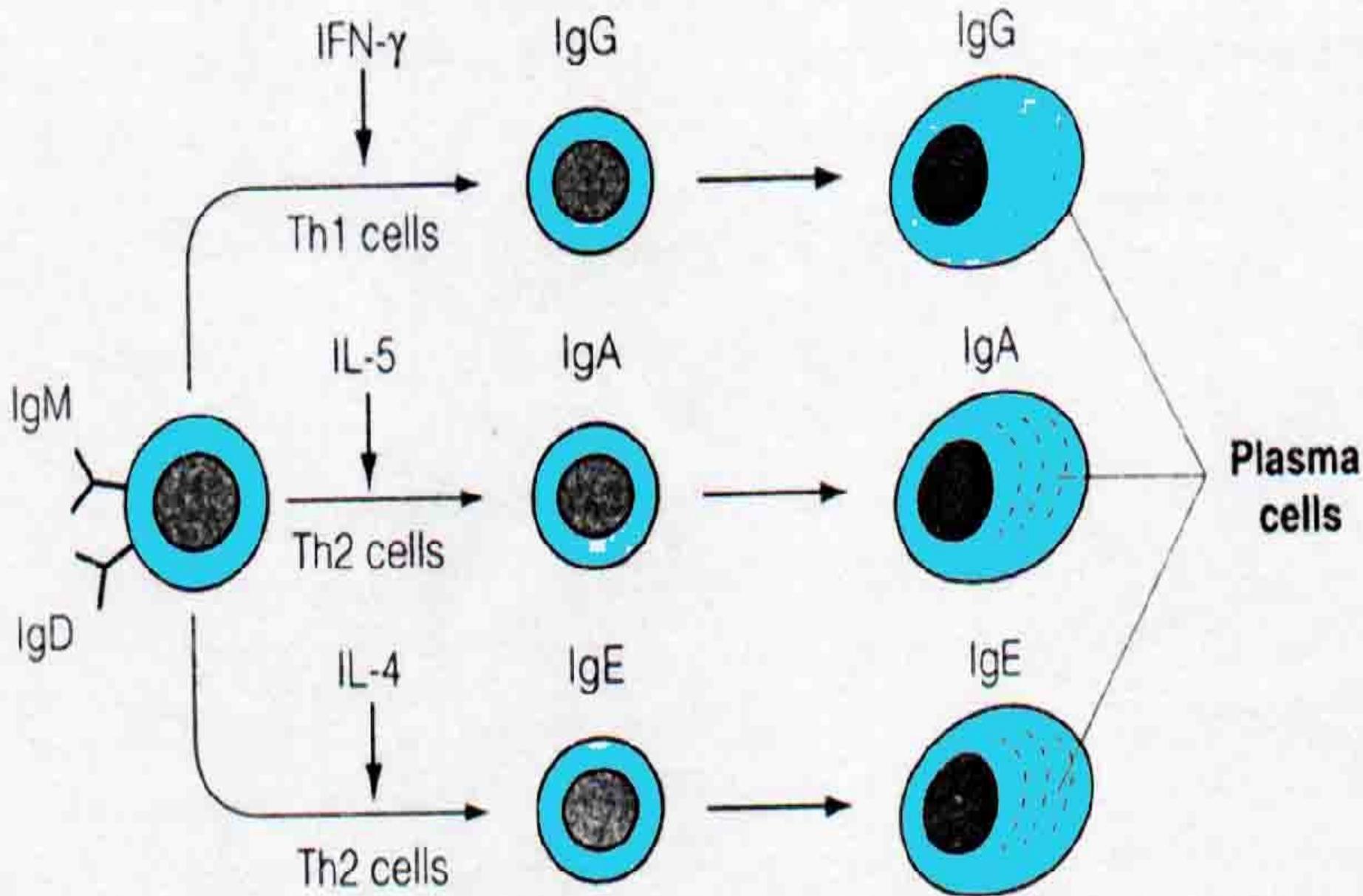
Secondary Lymphoid Organs

ACTIVATION OF B CELL

- ✓ Activation of B cell leads to proliferation and differentiation into a plasma cell, the end stage of B cell differentiation.
- ✓ It occurs in specialized regions of secondary lymphoid organs (spleen, Lymphoid nodes), the germinal centers.
- ✓ B cell gets activated by:
 - ✓ Antigen
 - ✓ Class or Isotype switching.





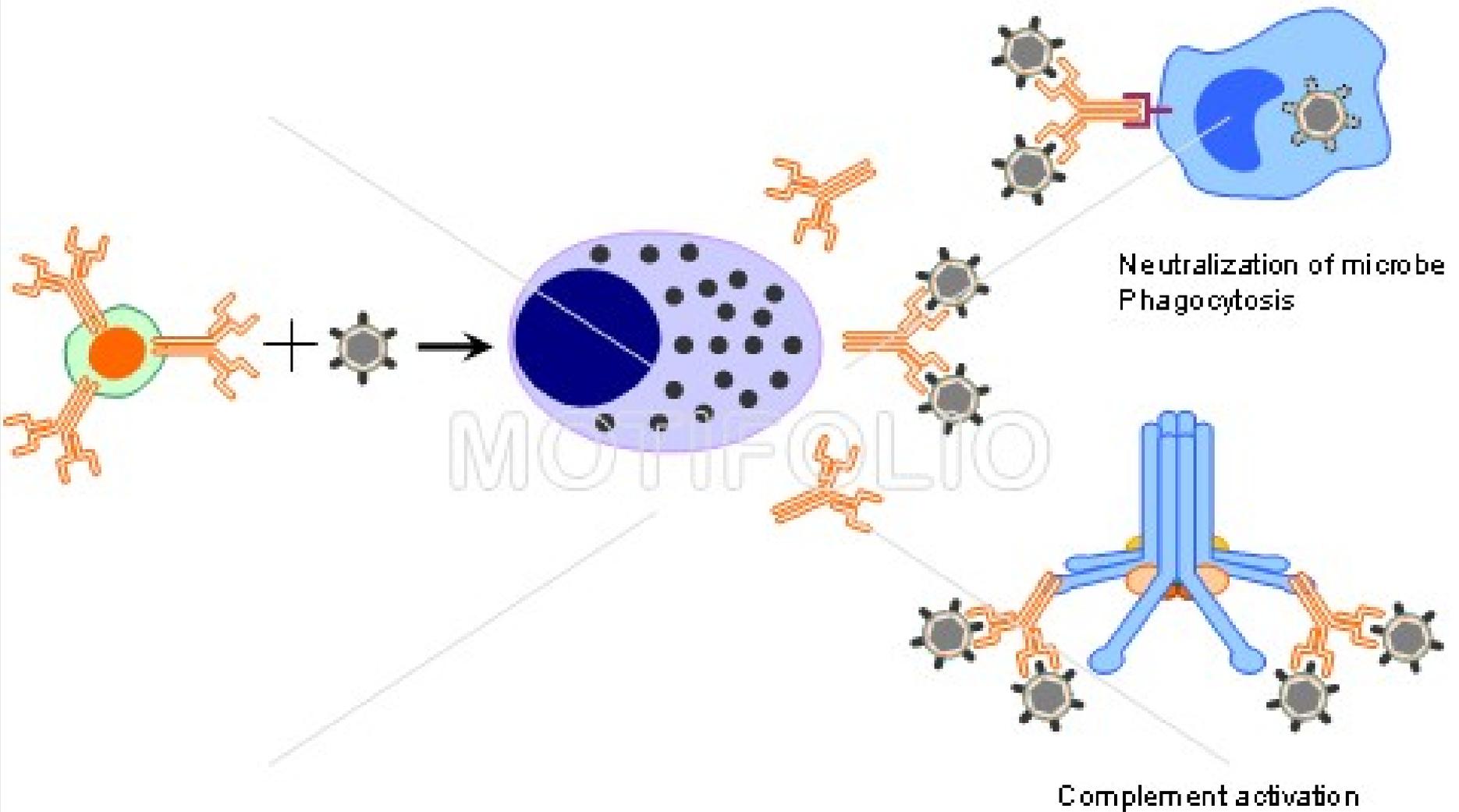


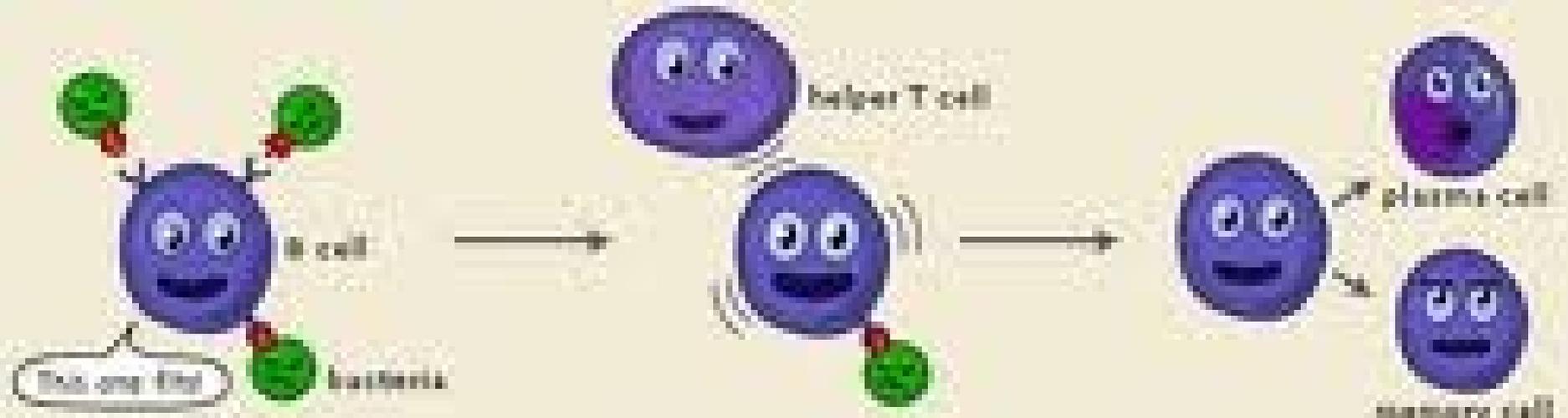
FUNCTION OF B CELLS

- ✓ To interact with antigenic epitopes, using their immunoglobulin receptors
- ✓ To present antigenic peptides to T cells, consequent upon interiorization and processing of the original antigen.
- ✓ As the B cells are responsible for the humoral arm of the adaptive immune system, it act against extracellular pathogens.
- ✓ Ig when bound to the cognate antigens, can activate the complement system and help phagocytes to take up antigens.



B lymphocyte and it's function





1. The B cell finds an antigen which matches its receptors.
2. It waits until it is activated by a helper T cell.
3. Then the B cell divides to produce plasma and memory cells.



4. Plasma cells produce antibodies that attach to the current type of invader.
5. "Eater cells," prefer intruders marked with antibodies, and "eat" loads of them.
6. If the same intruder invades again, memory cells help the immune system to activate much faster.

HUMORAL/B CELL DEFECT

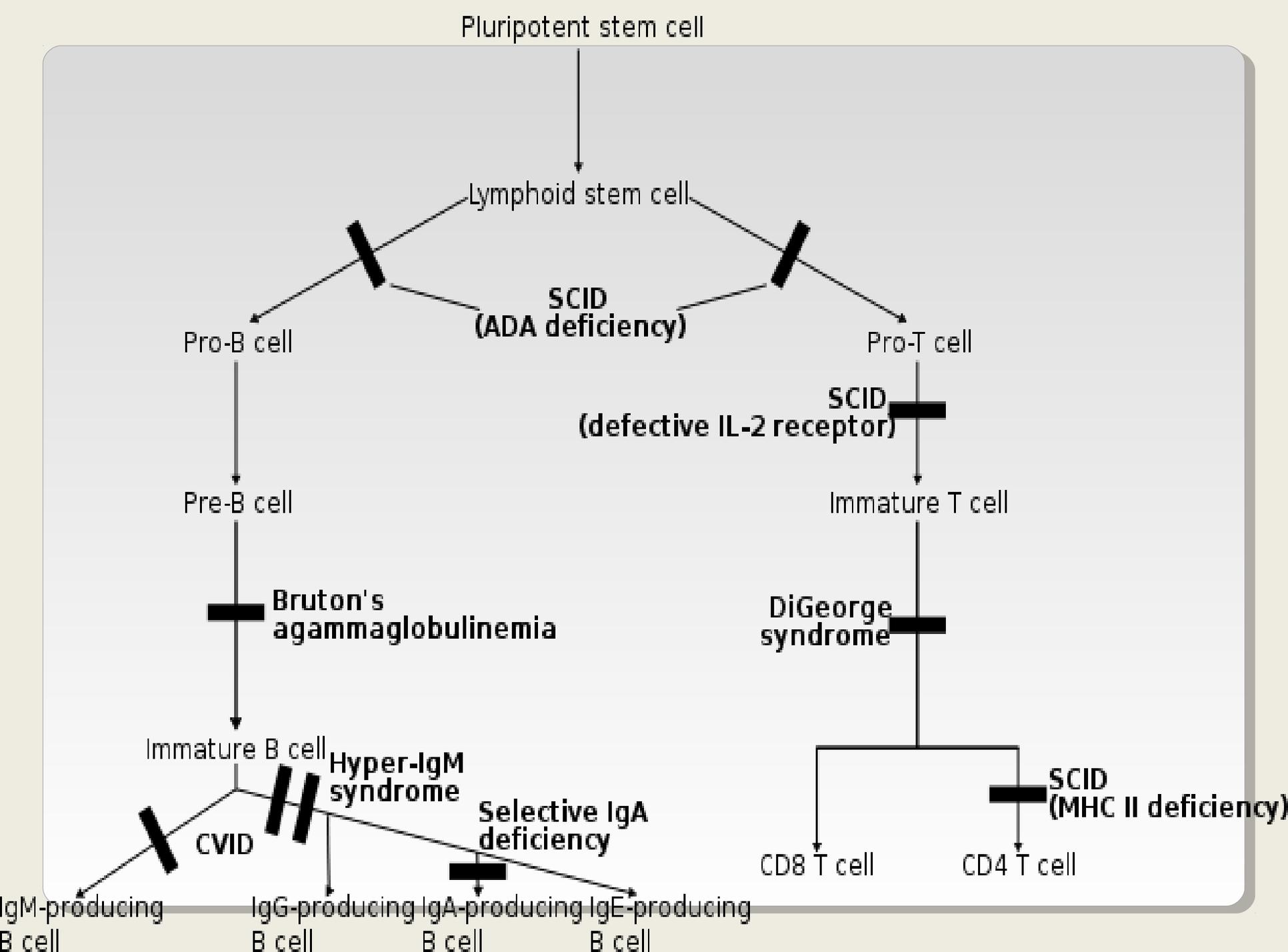


B CELL IMMUNODEFICIENCY

- What does it mean B cell defect?
 - ✓ Defects in antibody production
- As a result there will be:
 - ✓ poor opsonization of bacteria
 - ✓ recurrent infection with encapsulated bacteria

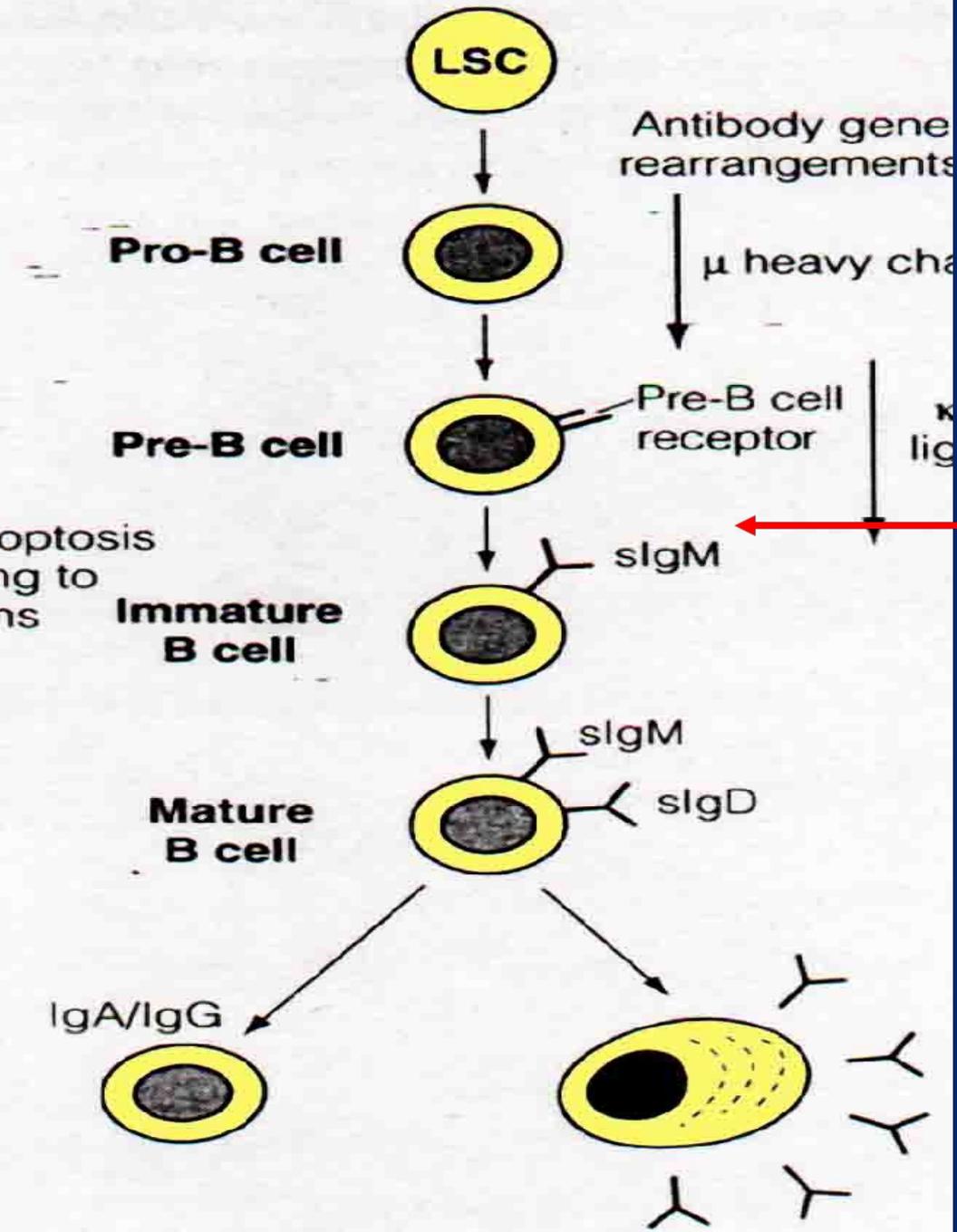
DISEASES CAUSED BY B CELL DEFECT

- B cell deficiency leads to different types of diseases they are:
 - ✓ X-linked agammaglobulinemia (X-LA)
 - ✓ IgA deficiency
 - ✓ IgG subclass deficiency
 - ✓ immunodeficiency with increased IgM (HIgM)
 - ✓ common variable immunodeficiency (CVID)
 - ✓ transient hypogammaglobulinemia of infancy



X-LINKED HYPOGAMMAGLOBULINEMIA

- Causes of the disease:
 - ✓ Mutation in the gene called *bruton thyroid kinase(BTK)* which *encodes tyrosine kinase*, an important signal transduction protein.
- Diagnosis:
 - ✓ Virtual absence of B-cells
 - ✓ Very low level of all immunoglobulins(IgG, IgA, IgM, IgD and IgE)



Bruton disease

— mutations in *btk*

— maturation disorder
of pre-B cell

➤ Clinical Signs:

- ✓ Recurrent pyogenic bacterial infection.
- ✓ Eg: otitis media, sinusitis, respiratory infections.

➤ Treatment:

- ✓ Pooled gamma globulin infused intravenously.



ISOLATED IGA DEFICIENCY

- Heterogeneous disorder, Most common immunodeficiency
- Causes:
 - ✓ Unknown
 - ✓ Associated with a decreased release of IgA synthesized by B lymphocytes
- Clinical manifestation:
 - ✓ Recurrent sinopulmonary viral or bacterial infection.
 - ✓ GI tract infection.
 - ✓ Tend to develop immune complex diseases(type III hypersensitivity)
 - ✓ Paradoxically, most people with IgA deficiency remain generally healthy.
- Treatment:
 - Administration of wide-spectrum antibodies

X-LINKED IMMUNODEFICIENCY WITH HYPER-IGM (XHM)

➤ Causes:

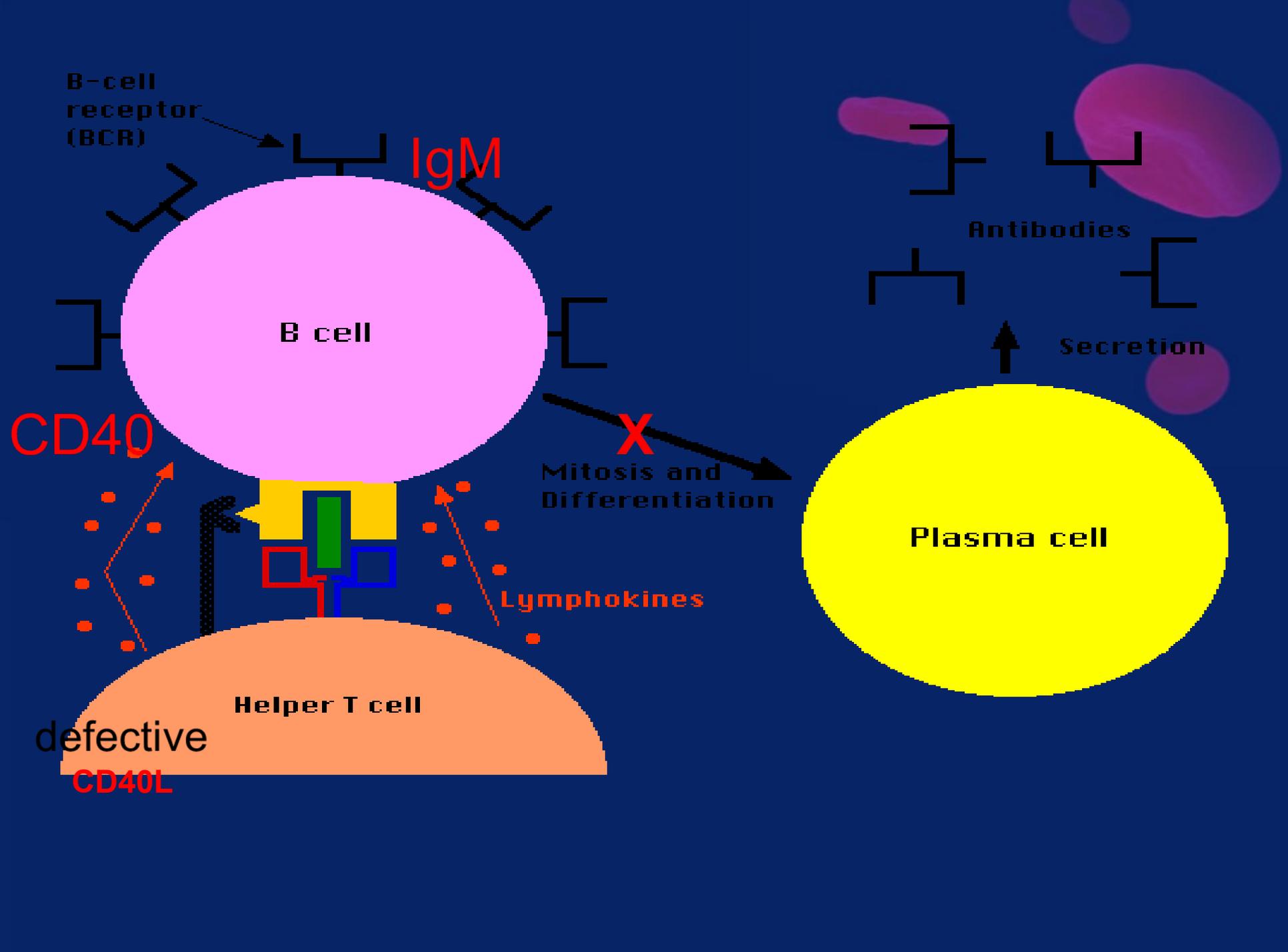
- is related to a deficiency in gp39 (CD40 ligand)
- No isotype switching-results in a situations where individuals are IgG and IgA deficient, but synthesis large amounts of polyclonal IgM.

➤ Clinical Manifestations:

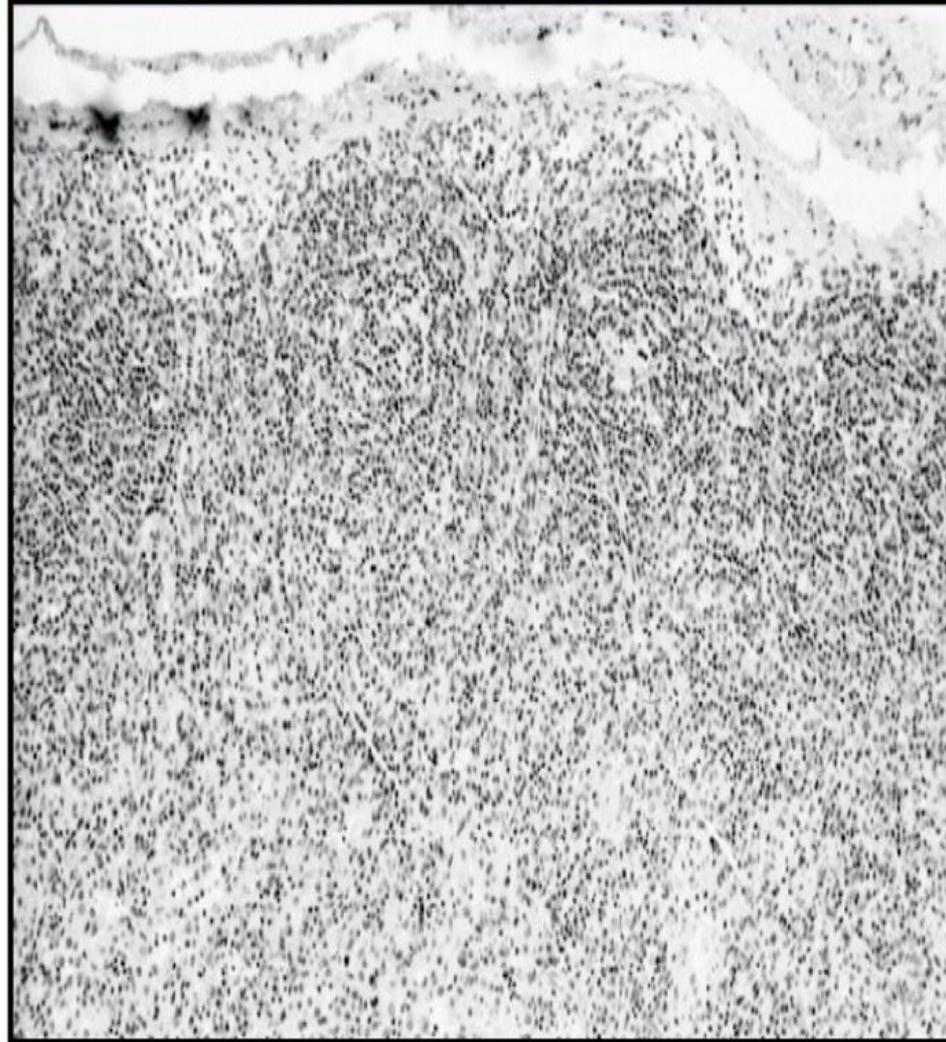
- Highly susceptible to pyogenic infection.
- Complexities of autoimmune disease
- No germial centers in lymph nodes.

➤ Treatment:

- Treated with intravenous gammaglobulins.



Lymph node from patient with hyper-IgM syndrome (no germinal centers)



Normal lymph node with germinal centers

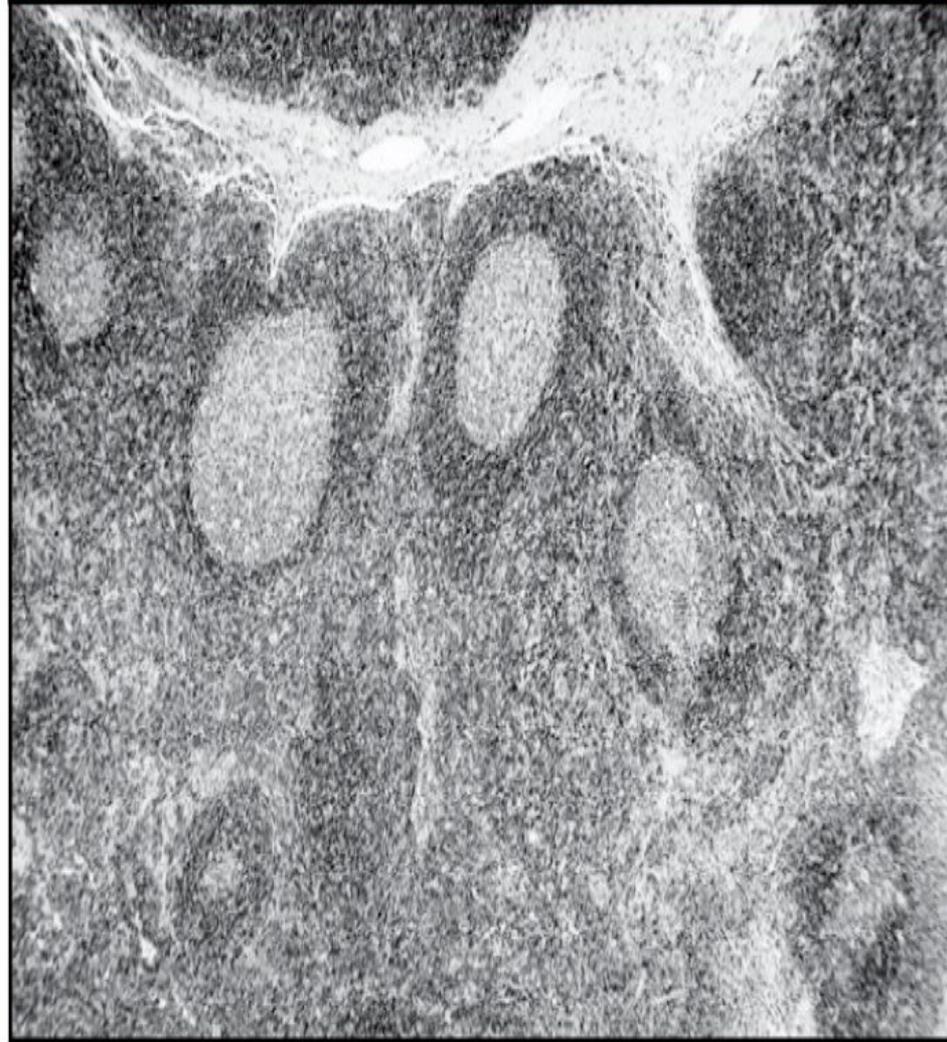


Figure 11-12 Immunobiology, 6/e. (© Garland Science 2005)

COMMON VARIABLE IMMUNODEFICIENCY

➤ Causes:

- ✓ mature B cells are normal in number and morphology but fail to differentiate to plasma cells because of defective interaction between T and B cells, mostly caused by a T-cell defect.
- ✓ This defect is thought to be related to a decreased number and/or function of CD4⁺ T lymphocytes or, occasionally, to an increased number of CD8⁺ T lymphocytes.

➤ Clinical manifestation:

- ✓ Pyogenic bacteria, viruses such as Epstein-Barr virus, Giardia lamblia (protozoan).
- ✓ High incidence of autoimmune diseases such as hemolytic anemia, thrombocytopenia, SLE.

➤ Treatment:

- ✓ Intravenous gammaglobulin must be given for the protection.

Warning signs for B cell immunodeficiency

Medical history

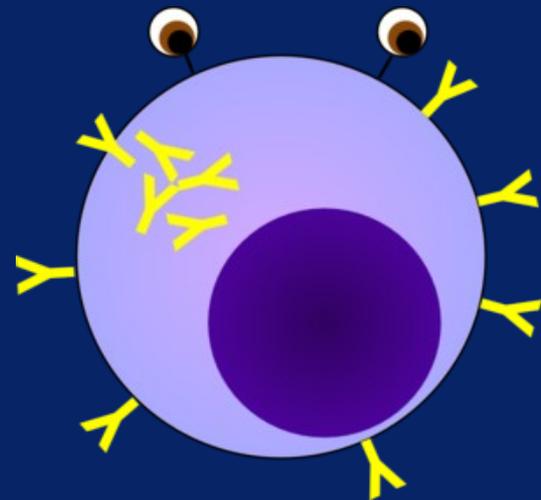
- ≥ 8 ear infections / year
- ≥ 2 serious sinusitis / year
- ≥ 2 pneumonias / year
- ≥ 2 deep-seated infections, or infections in unusual areas
- Recurrent deep skin/organ abscesses
- Need for IV ABx to clear infection
- Infections with unusual /opportunistic organisms
- Family Hx of primary immunodeficiency

Physical signs

- Poor growth, FTT
- Absent lymph nodes or tonsils
- Skin lesions: telangiectasias, petechiae, lupus-like rash
- Ataxia (ataxia-telangiectasia)
- Oral thrush after 1 year of age
- Oral ulcers

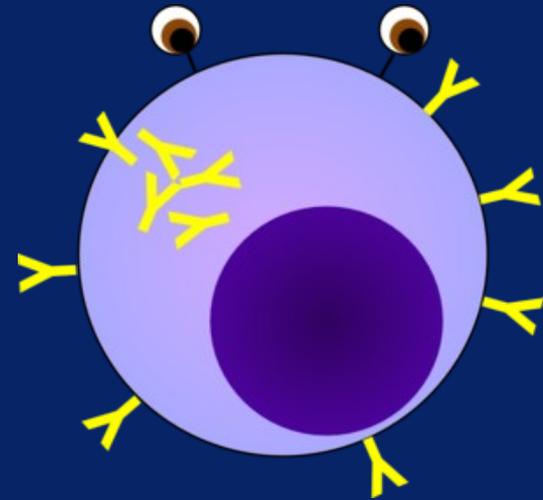
LABORATORY DIAGNOSIS

- *Blood count..*
- *Quantitative immunoglobulins.*
- *Antibody responses.*
- *Complement.*
- *Skin tests.*

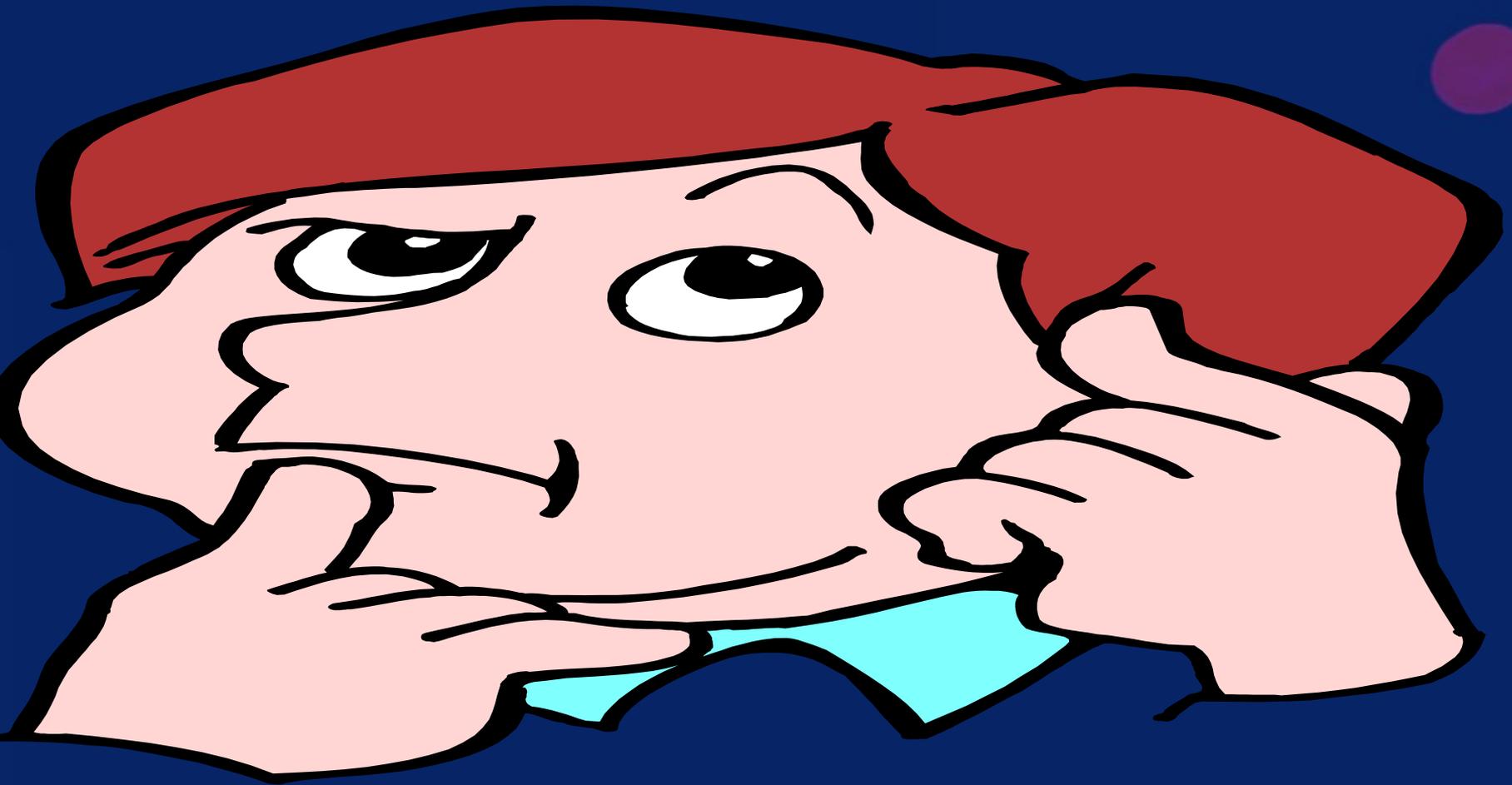
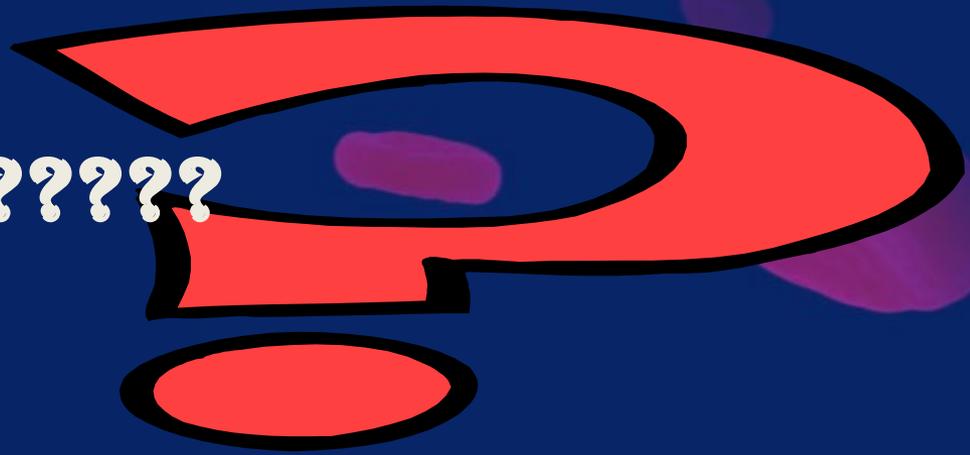


TREATMENT

- ✓ Gammaglobulin infusion thru Intravenous
- ✓ Gene replacement therapy
- ✓ Antibiotics for bacterial infections and other primary infections.



QUESTIONS PLS??????





Thank
you!

