Immune deficiency? How to Assess

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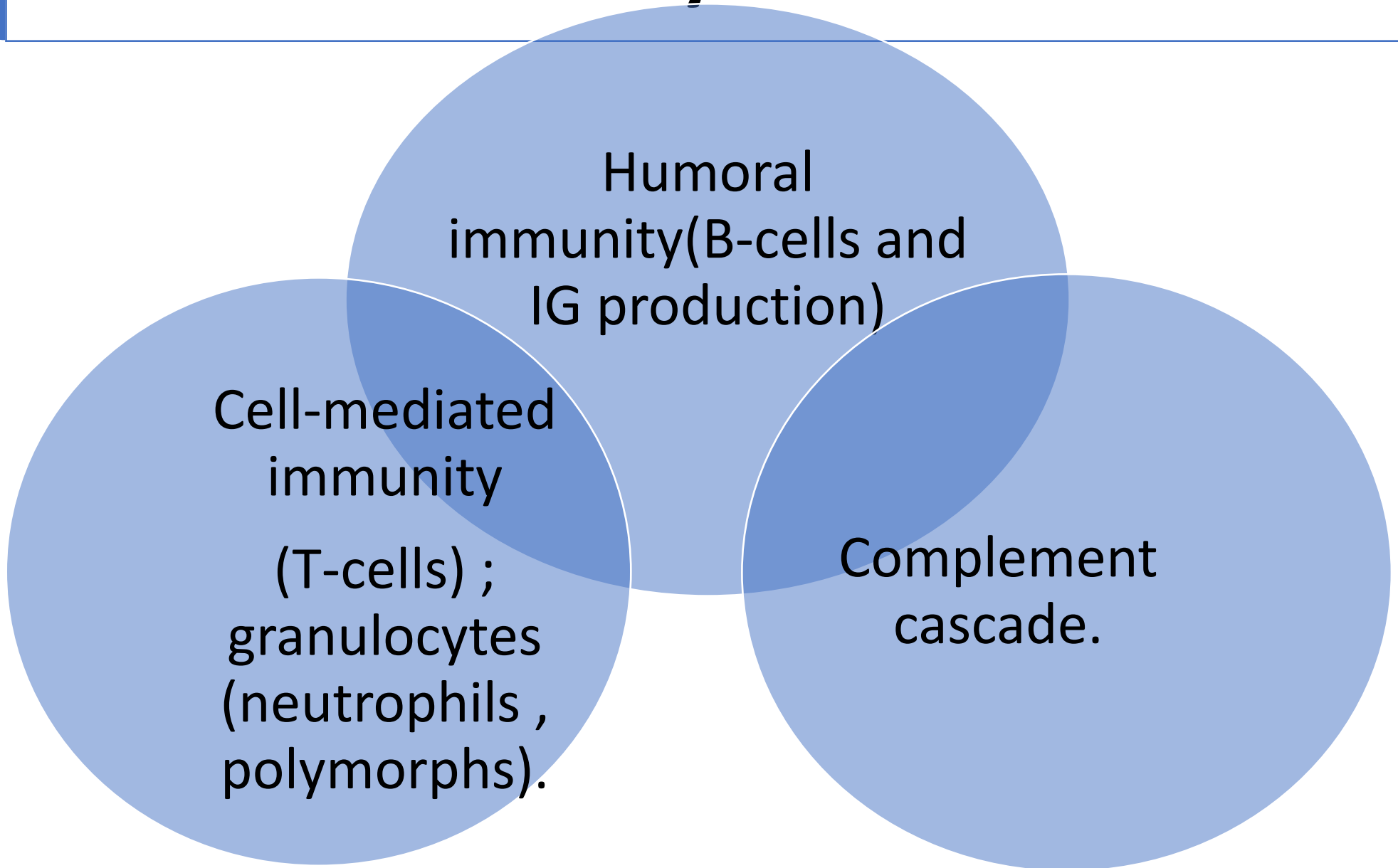
Introduction

Definition:
Immunodeficiency is the absence or failure of normal function of one or more elements of the immune system.

Results in immunodeficiency disease:

- Recurrent infection.
- Autoimmunity as a result of immune dysregulation.
- Increased susceptibility to malignancy.

Immunity to infection



Classification

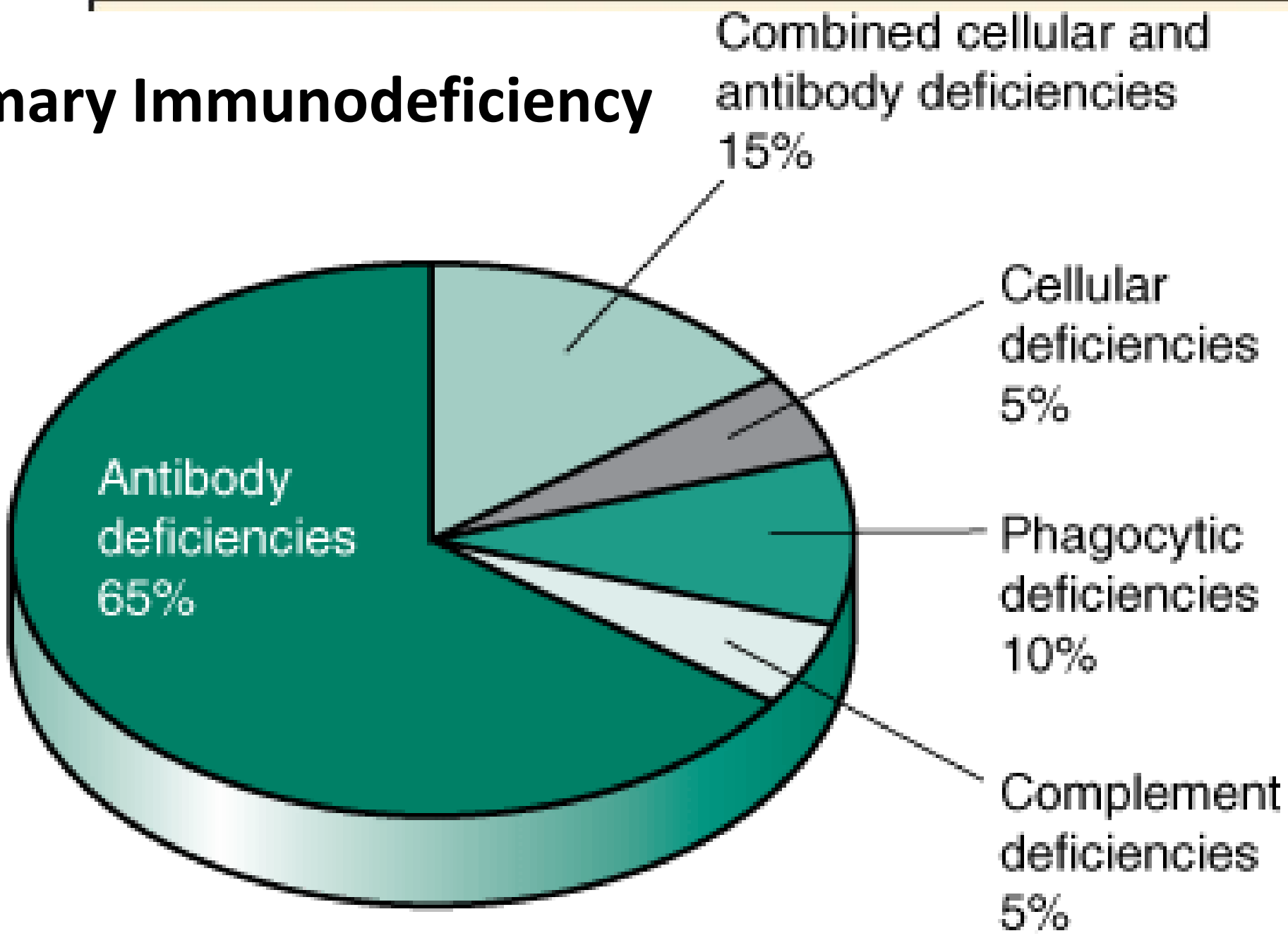
Primary Immunodeficiency:

- Usually congenital, resulting from genetic defects in some components of the immune system.

Secondary (Acquired):

- As a result of other diseases or conditions.

Primary Immunodeficiency



Evaluation: Step 1: Warning signs of immunodeficiencies?

- ≥ 4 ear infections in one year.
- ≥ 2 severe sinus infections in one year.
- ≥ 2 months treatment of antibiotics with little effect.
- ≥ 2 pneumonias per year.
- Insufficient weight gain or growth delay.
- Recurrent deep skin or organ abscesses (ex. liver, lungs)
- Persistent thrush in mouth or fungal infection on skin.
- Need for intravenous antibiotics to clear infections.
- ≥ 2 deep seated infections (ex. septicemia, meningitis)
- Family history of a primary immunodeficiency.

Step 2: Rule out common causes of infection and secondary immunodeficiency.

- **Common causes of infection:**
  - **Recurrent otitis media (**Eustachian tube dysfunction due to allergy)
  - **Sinusitis (**Nasal polyps, Allergic rhinitis)
  - **Pneumonia (**Recurrent aspiration (alcohol/drug use, seizure disorder), GERD)
- **Secondary immunodeficiency:**
  - DM, CKD, Malnutrition, HIV, Drugs.

Step 3: Consider primary immunodeficiencies-Categorize considering types and locations of infections, age of patient and other associated findings.

- Recurrent and severe sinopulmonary infections with encapsulated bacterial strains (eg, Streptococcus pneumoniae, Haemophilus influenzae)→Antibody deficiency.
- Severe viral and/or bacterial illnesses or opportunistic infections → cellular immunity defect (T Cell).
- Recurrent Neisserial infections →Complement defect.
- Recurrent abscesses in unusual site → Phagocytic defect.
- Early Presentation (< 6 months) →SCID.

Step 4: Categorize the patient and order screening tests.

Step 5: Consider referral to immunology specialist and secondary lab tests.

Laboratory tests for immune system

Initial screening laboratory tests:

CBC: PBF:

- Neutropenia.
- Lymphopenia.
- Leukocytosis: If persistently elevated ANC, LAD is suspected.

Laboratory tests: (cont..)

RBS:

Renal function:

Albumin, globulin (low serum proteins suggest malnutrition or protein loss, Markedly elevated globulin levels may be seen in gammopathies or chronic infections).

Tests to evaluate for specific infections, if indicated by the presentation (e.g., appropriate cultures, chest and/or sinus imaging):

Tests for T- cell defect

- Absolute lymphocyte count.
- Flow cytometry

Tests for B cell defect

- IgA, IgM, IgG.
- IgE
- Antibody titer to protein & polysaccharides antigens (e.g. Pneumococcal vaccine)

Tests for phagocytic cell disorder

- Absolute neutrophil count.
- Respiratory burst assay.

Tests for complement deficiency

- CH50- If abnormal, individual complement level.
- AH-50.

SUMMARY

Step 1: "Too many" infections. (warning signs).

Step 2: Rule out common causes of infection (asthma, foreign body) and secondary immunodeficiency (malnutrition, HIV).

Step 3: Consider primary immunodeficiencies- Categorize by types and locations of infections, age of patient.

Step 4: Categorize the patient and order screening tests.

Antibody deficiency (B lymphocyte)	Cellular immunity Deficiency (T lymphocyte)	Phagocyte deficiency	Complement deficiency

Step 5: Consider referral to immunology specialist and secondary lab tests.


Thank you