Primary Immunodeficiency

Definition: Inborn errors affecting immune factors, leading to deficient immunity.

Categories: Congenital immunodeficiency. Approximately 50% due to B cell issues. Around 30% linked to T cell defects. Roughly 18% are errors in phagocytes. Approximately 2% relate to complement deficiencies

Secondary Immunodeficiency

Definition: Acquired immunodeficiencies, more common than primary immunodeficiencies.

Causes: Age. Certain infectious agents. Medical interventions. Systemic disorders (e.g., diabetes, malnutrition, alcoholism, hepatitis).

Autoimmune Disorders and Diagnosing

Autoimmunity: Immune system attacks healthy self-tissues. Genetics and exposure to certain infectious agents contribute. Over 100 autoimmune diseases described.

Diagnosis: General signs and symptoms. Presentation varies based on affected tissues. Hypersensitivities are inappropriate immune responses. More common in developed nations (hygiene hypothesis).

Example of Primary Immunodeficiency

DiGeorge Syndrome

Deletion in part of chromosome 22. Causes impaired thymus development. Impairs cellular immune responses.

intervetions secondary Immunodeficiencies

Cancer treatments (e.g., radiation and chemotherapy). Steroid anti-inflammatory drugs (e.g., corticosteroids). Anti-seizure medications.

Allergy and Type I Hypersensitivities

Allergy: Triggered by allergens, leading to IgE production. Immune system reacts to harmless substances. Examples: atopic asthma, atopic dermatitis.

Systemic Anaphylaxis: Localized and systemic anaphylaxis. Systemic anaphylaxis is potentially life-threatening and treated with epinephrine.

Diagnosis and Management: Diagnosis based on symptoms, blood/skin tests. Management includes allergen avoidance, medications, and desensitization immunotherapy.

Type 4 Hypersensitivities/autoimmune

Type IV Hypersensitivities: T cell-mediated, not antibody-mediated. Manifest slowly, causing delayed hypersensitivity reactions. Responsible for autoimmune disorders.

Autoimmune Type IV Hypersensitivities: Examples: Guillain-Barré syndrome, Hashimoto thyroiditis, Type I diabetes, multiple sclerosis, celiac disease. Therapies aim to reduce T cell response and inflammation.

Nonautoimmune Type IV Hypersensitivities: Triggered by haptens. Examples: tuberculin skin test, contact dermatitis, transplant rejection, graft-versus-host disease.

Therapies for Primary Immunodeficiencies

Bone marrow transplants. Intravenous or subcutaneous antibody administration. Cytokine therapies. Experimental treatments (e.g., stem cell transplants, thymus transplantation, gene therapy).

Immune Deficiencies and Cancer

The immune system protects against infections and cancer.

Patients with compromised immune systems are at increased risk for certain cancers.

Immunotherapies aim to boost immune defenses to prevent or treat cancer.

Type II and Type III Hypersensitivities

Type II Hypersensitivities: IgG or IgM bind to cell surface or extracellular antigens, leading to complement activation and cell lysis. Examples: Goodpasture syndrome, autoimmune hemolytic anemia, rheumatic heart disease.

Blood Groups and Transfusion Reactions: Blood types based on antigens on red blood cells. Incompatible transfusions cause hemolytic reactions.

Type III Hypersensitivities: IgG or IgM antibodies bind to soluble targets, forming immune complexes. Examples: antivenoms, antitoxins.

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