Auto -immune Disorder in the Children

Done by: Mariam Mohammad

MSc. Pediatric Nursing

Outline:

- Introduction
- Anatomy and physiology of the immune system
- Assessment of immune system
- Causes Autoimmune Disease.
- Symptoms of Autoimmune Disease
- Diagnosing Autoimmune Disease
- Systemic Lupus Erythematosus (SLE).
- Crohn ' s Disease

Autoimmune Disease

Introduction:

The immune system consists of specialized cells and organs that work together to protect an organism from outside influences.

It operates throughout the body, but certain sites contain cells organized for specific functions. When the immune system is functioning properly, it protects the body against bacteria, viral infections, cancer cells, and other substances that are foreign to the organism.

If the immune system weakens, the body's ability to defend itself also weakens, and pathogens can develop. These are infectious agents that can cause disease or illness to its host.

Anatomy of the immune system:-

There are fluids and Organs are responsible about the immunity.

Two main fluid systems in the body:-

- Blood
- Lymph

Are intertwined throughout the body and they are responsible for transporting the agents of the immune system.

Fluid Systems of the Body

1. The Blood System:

- Hemocytoblasts mature into three types of blood cells: ***erythrocytes*** (red blood cells or RBCs), ***leukocytes*** (white blood cells or WBCs), and ***thrombocytes*** (platelets).
- The leukocytes are further subdivided into **granulocytes** (containing large granules in the cytoplasm) and **agranulocytes** (without granules). The granulocytes consist of ***neutrophils*** (55–70%), ***Eosinophils*** (1–3%), and ***basophiles*** (0.5–1.0%).
- The agranulocytes are ***lymphocytes*** (consisting of B cells and T cells) and ***monocytes***. Lymphocytes circulate in the blood and lymph systems, and make their home in the lymphoid organs.
- There are 5000–10,000 WBCs per mm^3 and they live 5-9 days.

2. The Lymph System:

- Lymph is an alkaline (pH > 7.0) fluid that is usually clear, transparent, and colorless. It flows in the lymphatic vessels and bathes tissues and organs in its protective covering. There are no RBCs in lymph and it has lower protein content than blood. Like blood, it is slightly heavier than water (density = 1.019 ± .003).
- The lymph flows from the interstitial fluid through lymphatic vessels up to either:-
    - The thoracic duct or
    - Right lymph duct, which terminate in the subclavian veins, where lymph is mixed into the blood. (The right lymph duct drains the right sides of the thorax, neck, and head, whereas the thoracic duct drains the rest of the body).

- ❖ Lymph carries lipids and lipid-soluble vitamins absorbed from the gastrointestinal tract (GIT). Since there is no active pump in the lymph system, there is no back-pressure produced.
- ❖ The lymphatic vessels, like veins, have one-way valves that prevent backflow. Additionally, along these vessels there are small bean-shaped ***lymph nodes*** that serve as filters of the lymphatic fluid. In this lymph nodes antigen is usually presented to the immune system.
- ❖ Lymphoid tissue is a very important piece of the defenses of the immune system because it:
  1. Houses and provides a proliferation site for lymphocytes.
  2. creates a surveillance checkpoint for lymphocytes and macrophages.

Organs of Immunity System:-

The human **lymphoid system** has the following:

1. Primary organs: bone marrow (in the hollow center of bones) and the thymus gland (located behind the breastbone above the heart):

a) Bone marrow:

All the cells of the immune system are derived from stem cells in the bone marrow. The bone marrow is the site of origin of red blood cells, white cells (including lymphocytes and macrophages) and platelets.

b) Thymus:

- ❖ In the thymus gland lymphoid cells undergo a process of maturation and education prior to release into the circulation. This process allows T cells to develop the important attribute known as self tolerance.
- ❖ The thymus gland is found in the thorax in the anterior mediastinum. It gradually enlarges during childhood but after puberty it undergoes a process of involution resulting in a reduction in the functioning mass of the gland. It continues to function throughout life, however.
- ❖ The thymus gland is arranged into an outer, more cellular, cortex, and an inner, less cellular, medulla. Immature lymphoid cells enter the cortex proliferate, mature, and pass on to the medulla. From the medulla mature T lymphocytes enter the circulation. Thymus releases hormones that enable T lymphocytes to function against specific pathogens.
- ❖ Differs from other lymphatic organs in that thymus does not directly fight antigens because it is only functioning with maturing T lymphocytes, and the structure is different from other organs.

2. Secondary organs: at or near possible portals of entry for pathogens: adenoids, tonsils, spleen (located at the upper left of the abdomen), lymph nodes (along the lymphatic vessels with concentrations in the neck, armpits, abdomen, and groin), Peyer's patches (within the intestines), and the appendix.

a) Tonsils:

It is the simplest lymphatic organs, which forms a ring of lymphatic tissue around the entrance to the throat; they gather and remove many of the pathogens in inhaled air of food.

- ❖ **Palatine tonsils**: located on either side of the posterior oral cavity; largest and most often infected.
- ❖ **Lingual tonsils**: Lies at the base of the tongue.
- ❖ **Pharyngeal tonsils**: are present in the posterior wall of the nasopharynx.
- ❖ **Tubal tonsils:** surround the openings of the auditory tubes into the pharynx.

b) <u>Spleen:</u>

- ❖ The spleen is located in the upper left quadrant of the abdomen. It is about the size of a fist and is the largest lymphatic organ. It has two main functions acting as part of the immune system and as a filter.
- ❖ The spleen acts as a blood filter removing defective and old blood cells, debris, foreign matter, bacteria, viruses and toxins. It also provides the advantages described by having lymphatic tissue. Also it stores breakdown particles from blood cells for later use, stores blood platelets, and is the site of erythrocyte production in the fetus.
- ❖ There are **two** distinct components of the spleen, the **red pulp** and the **white pulp**.

- Red pulp: There is a complex system of blood vessels within the red pulp arranged to facilitate removal of old or damaged red blood cells from the circulation.

- White Pulp: The white pulp contains T cells, B cells and accessory cells. There are many similarities with lymph node structure. The purpose of the white pulp is to mount an immunological response to antigens within the blood. The white pulp is present in the form of a periarteriolar lymphoid sheath. This sheath contains B cell follicles and T cells. At the edge of the T zone is a region known as the marginal zone where larger lymphocytes and antigen presenting dendritic cells are located.

c) <u>Lymph nodes</u>

- ❖ Lymph nodes are small bean shaped structures lying along the course of lymphatics. They are aggregated in particular sites such as the neck, axillae, groins and para-aortic region.
- ❖ The node is made up of **three components**:
  - Lymphatic sinuses.
  - Blood vessels.
  - Parenchyma (cortex, paracortex, medulla).
- ❖ Lymph nodes **have two key roles in body defense**:-

1. They act as lymph filters, where macrophages in the nodes remove and destroy microorganisms, preventing them from attacking the body.

2. They help to activate the immune system. Lymphocytes in the node monitor the lymph stream for antigens and attack them when they are found.

d) Peyer's Patches:

❖ These are quite large aggregates of lymphoid tissue found in the small intestine. The overlying 'dome' epithelium contains large numbers of intraepithelial lymphocytes. Peyer's Patches facilitate the generation of an immune response within the mucosa. B cell precursors and memory cells are stimulated by antigen in Peyer's Patches. Cells pass to the mesenteric lymph nodes where the immune response is amplified. Activated lymphocytes pass into the blood stream via the thoracic duct.

❖ Peyer's patches help protect the body against bacterial invasion through k lymphocytes present in intestinal of intestinal tract through the intestinal wall.

Physiology of the immune system

The immune system is usually divided into two sections,

- innate immunity and
- Adaptive immunity.

A. **Innate immunity:-** is a set of encoded receptors to pathogens that do not change during the lifetime of the organism. It is developed by hereditary components and helps to immediately defend and get rid of pathogens.

B. **Adaptive immunity**:- consists of responses to pathogens changing and developing during the lifetime of the organism.

Immunity can be initiate or adaptive.

Adaptive immunity is divided:-

❖ according to its **source** to natural and artificial immunity,.

❖ according to its **mechanisms** to humoral immunity and cell-mediated immunity.

A. Innate Immunity

The innate immunity system is what we are born with and it is nonspecific; all antigens are attacked pretty much equally. It is genetically based and we pass it on to our offspring.

1. Skin and Mucosa:

- This is the body's first line of defense against attack from harmful microorganisms.
- Highly effective when unbroken, the keratinized skin forms a tough defense against outside threats.

- Mucous membranes also form physical barriers, and they both produce defensive chemicals.
- Acidity of skin secretions inhibits bacterial growth.
- The stomach secretes hydrochloric acid and protein digesting enzymes which both kill pathogens.
- Saliva and lacrimal fluid contain lysozyme, which is an enzyme that kills bacteria.
- The sticky mucous also traps many particles that otherwise would have entered the body.

2. Phagocytes:

- A phagocyte is a cell that attracts (by chemotaxis), adheres to, engulfs, and ingests foreign bodies.
- Phagocytes including granulocytes, monocytes (white blood cells) and specialized tissue cells (macrophages) are major cellular components of the non-specific immune system.
- Phagocytosis is the process by which pathogens are engulfed and destroyed by phagocytes of the host's immune system.
- Macrophages travel throughout the body looking for invading particles that could harm the body –called free macrophages.
- Fixed macrophages also occur in the liver and lungs.
- Macrophages derived from white blood cells that leave the blood stream.
- Phagocytes use similar process to that of an amoeba in that they engulf the invading particle and digest it.

3. Natural Killer Cells:

- They are large granular lymphocytes that attach to the glycoproteins on the surfaces of infected cells and kill them.
- Police the body and can attack and destroy cancer and virus infected cells before the immune system is activated.
- Present in blood and lymph.
- They can react to any virus infected or tumor cells, not just specific kinds.
- They don’t engulf like phagocytes; they attack the targets membrane with chemicals that soon cause the nucleus to disintegrate.

4. The complement system:

- It is a major triggered enzyme plasma system. It coats microbes with molecules that make them more susceptible to engulfment by phagocytes.
- Vascular permeability mediators increase the permeability of the capillaries to allow more plasma and complement fluid to flow to the site of infection.
- They also encourage polys to adhere to the walls of capillaries (margination) from which they can squeeze through in a matter of minutes to arrive at a damaged area.
- Once phagocytes do their job, they die and their "corpses," pockets of damaged tissue, and fluid form pus.

5. Eosinophils:

- They attracted to cells coated with complement C3B, where they release major basic protein (MBP), cationic protein, and oxygen metabolites, all of which work together to burn holes in cells and helminthes (worms).
- About 13% of the WBCs are eosinophils. Their lifespan is about 8–12 days. Neutrophils, eosinophils, and macrophages are all phagocytes.

6. Dendritic cells:

- They are covered with a maze of membranous processes that look like nerve cell dendrites. Most of them are highly efficient antigen presenting cells.
- There are four basic types: Langerhans cells, interstitial dendritic cells, interdigitating dendritic cells, and circulating dendritic cells.
- Langerhans cells are found in the epidermis and mucous membranes, especially in the anal, vaginal, and oral cavities. These cells make a point of attracting antigen and efficiently presenting it to T helper cells for their activation. [This accounts, in part, for the transmission of HIV via sexual contact.]

7. Normal flora:

- They are the microbes, mostly bacteria, which live in and on the body with, usually, no harmful effects to us.
- We have about 1013 cells in our bodies and 1014 bacteria, most of which live in the large intestine then on skin also various bacteria live in the nose and mouth, while Lactobacilli live in the stomach and small intestine.
- Most of the bacteria in large bowel are anaerobes. The urogenitary tract is lightly colonized by various bacteria and diphtheroids. After puberty, the vagina is colonized by Lactobacillus aerophilus that ferment glycogen to maintain an acid pH.
- Normal flora fill almost all of the available ecological niches in the body and produce bacteriocidins, defensins, cationic proteins, and lactoferrin all of which work to destroy other bacteria that compete for their niche in the body.
- The resident bacteria can become problematic when they invade spaces in which they were not meant to be.

8. Inflammation:

- This is how tissue responds to injury.
- Prevents damaging agents from spreading.
- Disposes of cell debris and pathogens.
- Sets the stage for the repair process.
- Four cardinal signs of short term inflammation:
  - Redness.
  - Heat.
  - Swelling.
  - Pain.
  - Impairment of function considered fifth sign.

9. Fever:

- Widespread response to an infection.
- Abnormally high temperature caused by a response to pyrogens.
- High fevers are dangerous, but mild fever benefits body in defense.
- During fever, liver and spleen hold iron and zinc which bacteria need to reproduce
- It also increases metabolic rate of tissue cells which increase repair and defense processes.

B. Adaptive Immunity

- As we said before there are two fundamental adaptive mechanisms: **cell-mediated immunity** and **humoral immunity**.
- The responsible about adaptive immunity is Lymphocytes which come in two major types: B cells and T cells.
- Lymphocytes constitute 20–40% of the body's WBCs. Their total mass is about the same as that of the brain or liver.
- The peripheral blood contains 20–50% of circulating lymphocytes; the rest move in the lymph system.
- Roughly 80% of them are T cells, 15% B cells and remainder are null or undifferentiated cells.
- B cells are produced in the stem cells of the bone marrow; they produce antibody and oversee humoral immunity.
- T cells are nonantibody-producing lymphocytes which are also produced in the bone marrow but sensitized in the thymus and constitute the basis of cell-mediated immunity.

1. **Cell-mediated immunity:**

- Macrophages engulf antigens, process them internally, and then display parts of them on their surface together with some of their own proteins. This sensitizes the T cells to recognize these antigens.
- All cells are coated with various substances. CD stands for cluster of differentiation and there are more than one hundred and sixty clusters, each of which is a different chemical molecule that coats the surface. Every T and B cell has about 105 = 100,000 molecules on its surface.
- The large number of molecules on the surfaces of lymphocytes allows huge variability in the forms of the receptors.
- Cytotoxic or killer T cells (CD8+) do their work by releasing lymphotoxins, which cause cell lysis. Helper T cells (CD4+) serve as managers, directing the immune response. They secrete chemicals called lymphokines that stimulate cytotoxic T cells and B cells to grow and divide, attract neutrophils, and enhance the ability of macrophages to engulf and destroy microbes.
- Suppressor T cells inhibit the production of cytotoxic T cells once they are unneeded, lest they cause more damage than necessary. Memory T cells are

programmed to recognize and respond to a pathogen once it has invaded and been repelled.

2. Humoral immunity:

- An immunocompetent but as yet immature B-lymphocyte is stimulated to maturity when an antigen binds to its surface receptors and there is a T helper cell nearby (to release a cytokine).
- This sensitizes or primes the B cell and it undergoes clonal selection, which means it reproduces a sexually by mitosis. Most of the family of clones becomes plasma cells. These cells, after an initial lag, produce highly specific antibodies at a rate of as many as 2000 molecules per second for four to five days. The other B cells become long-lived memory cells.
- Antibodies, also called immunoglobulin or Igs, constitute the gamma globulin part of the blood proteins. They are soluble proteins secreted by the plasma offspring (clones) of primed B cells. The antibodies inactivate antigens by, (a) complement fixation (proteins attach to antigen surface and cause holes to form, i.e., cell lysis), (b) neutralization (binding to specific sites to prevent attachment—this is the same as taking their parking space), (c) agglutination (clumping), (d) precipitation (forcing insolubility and settling out of solution), and other more arcane methods.
- Constituents of gamma globulin are: IgG-76%, IgA-15%, IgM-8%, IgD-1%, and IgE-0.002% (responsible for autoimmune responses, such as allergies and diseases like arthritis, multiple sclerosis, and systemic lupus erythematosus).
- IgG is the only antibody that can cross the placental barrier to the fetus and it is responsible for the 3 to 6 month immune protection of newborns that is conferred by the mother.
- IgM is the dominant antibody produced in primary immune responses, while IgG dominates in secondary immune responses. IgM is physically much larger than the other immunoglobulins.

Assessment of immune system

I. Health history assessment.

II. Physical examination

I. Health history assessment

***Age:**

Extreme ages (old ages and infants) are at high risk to problem in their immune function.

***Nutrition:**

Good nutrition, fatty acid, lipids, vitamins, protein, trace element. All are essential for optimal function of immune system.

***Infection and immunization:**

- ❖ Recently and during childhood immunization.
- ❖ Present and past exposure to TB.
- ❖ Chest x ray.
- ❖ Recent exposure to any treatment.
- ❖ Fever of unknown origin.
- ❖ Lesion, sores, and ulcers.

***Allergy:**

- ❖ History of allergy (dust, paint, food).
- ❖ Symptoms experienced.
- ❖ Seasonal variation.
- ❖ Severity in symptoms.
- ❖ Treatment received and its effectiveness.
- ❖ Medications (Antihistamines, Corticosteroids, Epinephrine) and food allergies (milk, eggs, peanuts, soy, wheat, fish, shellfish such as shrimp).

***Food Allergy Reactions:-**

- Food allergy reactions can vary from person to person.
- Some reactions can be very mild and only involve one part of the body, like hives on the skin. Others can be more severe and involve more than one part of the body. Reactions can occur within a few minutes or up to a few hours after contact with the food.

Food allergy reactions can affect any of the four following areas of the body:

1. **skin:** itchy red bumps (hives); eczema; redness and swelling of the face or extremities; itching and swelling of the lips, tongue, or mouth (skin reactions are the most common type of reaction)
2. **gastrointestinal tract:** abdominal pain, nausea, vomiting, or diarrhea
3. **respiratory tract:** runny or stuffy nose, sneezing, coughing, wheezing, shortness of breath
4. **cardiovascular system:** lightheadedness or fainting

Types of Allergies:-

- **Cigarette Smoke Allergy** :-

Cigarette smoke allergy is caused by the numerous toxic chemicals and irritants found in cigarette smoke.

- **Dust Mite Allergy:-**

Dust mite allergy is an allergy to a microscopic organism that lives in the dust found in all dwellings.

- **Food Allergy :-**

A food allergy occurs when a food triggers an abnormal response by the body's immune system.

- **Grass Pollen Allergy**:-

Grass pollen allergy is a more difficult type of allergy to prevent because it is regional and seasonal.

- **Hay Fever :-**

Hay fever, also known as pollen allergy, is one of the most common allergies in the United States.

- **Mold Allergy :-**

A person can develop an allergy to the molds and yeasts that are commonly found indoors and outdoors.

- **Pet Allergy**:-

Many people believe pet allergies are triggered by the fur of cats and dogs. However, the major pet allergens are actually proteins in the saliva of animals.

- **Cockroach Allergy:-**

This common indoor pest allergy is caused by certain proteins found in cockroach saliva and feces

Skin Test for Allergy:-

- This is a test done on the skin to identify the allergy substance (the allergen) that is the trigger for an allergic reaction.
- A small amount of the suspected allergy-provoking substance (the allergen) is placed on the skin.
- The skin is then gently scratched through the small drop with a special sterile needle. An allergy skin test is also called a scratch test.
- If the skin reddens and, more importantly, if it swells, then the test is read as positive and allergy to that substance is considered probable.

Life style:-

Assess history of smoking, alcoholic, nutritional status and dietary intake, stress, occupation, exposure to radiation or pollution.

Immune disorders:-

Assess the history of having past and present exposure (as well as treatment and its effectiveness) for example:-

- Neoplastic disorders.
- Surgery.
- Chronic illness as D.M, renal diseases.
- Special problems (as burn, injury infection and stress)
- Medications (as antibiotics, corticosteroids and cytotoxic agents)
- Blood transfusion.

II. Physical examination

1 - Skin and mucus membrane assessment :-

for lesion, dermatitis, purpura, urticaria, inflammation, discharge, signs and symptoms of infection (skin temperature, presence of chills and sweating).

2- Anterior and posterior cervical, axillary and inguinal lymph nodes:

Palpate the lymph nodes for enlargement, location size and consistency.

3- Joints :-

for tenderness, swelling and limited range of motion.

4- Other body systems: Respiratory, cardiovascular, GIT, Genitourinary and neurosensory systems:-

for Signs and symptoms of immune dysfunction, level of stress and coping ability, function limitation.

Indications of immune dysfunctions

Respiratory system:

- Change in respiratory rate, cough.
- Abnormal lung sounds (wheezing, crackles).
- Rhinitis.
- Hyper ventilation and Bronchospasm.

Cardiovascular:

- Hypotension.
- Tachycardia.
- Dysrhythmias.
- Vasculitis.

- Anemia.

GIT:

- Hepatosplenomegaly.
- Colitis.
- Vomiting.
- Diarrhea.

Genitourinary system:

- Frequency.
- Burning urination.
- Hematuria.
- Discharge.

Neurosensory system:

- Cognitive dysfunction.
- Hearing loss.
- Visual changes.
- Headaches.
- Migraines.
- Ataxia.
- Tetany.

Autoimmune disorders fall into two general types:

- ✓ Systemic autoimmune diseases: damage many organs.
- ✓ Localized: only a single organ or tissue is directly damaged by the autoimmune process.

Autoimmune diseases that occur in children include:

- Lupus (SLE)
- Crohn ' s Disease
- Juvenile dermatomyositis
- Scleroderma
- Juvenile idiopathic arthritis (jia)
- Celiac disease

Causes of Autoimmune Disease:

- Environmental toxins.
- Heredity or genetics.
- Viruses.
- Sunlight.

- Iodine.
- Stress and anxiety.
- Pregnancy.
- Poor diet, lack of exercise, lack of sleep, abuse of alcohol and use of tobacco can also weaken the immune system.

Symptoms of Autoimmune Disease

The symptoms of autoimmune disease vary depending on the disease as well as the person's immune system. Common symptoms include:

- Anxiety, Irritability, or Depression.
- Hair Loss.
- Low or High Blood Pressure.
- Infertility or Low Sex Drive (Reduced Libido)
- Leg Cramps & Muscle Twitching.
- Acne.
- Dizziness.
- Sweating.
- Extreme sensitivity to cold in the hands and feet.
- Exhaustion & Fatigue.
- Nausea.
- Inflammation.
- Malaise.
- Elevated fever and High Body Temperature.
- Weakness and Stiffness in Muscles and Joints.
- Weight Changes.
- Digestive or Gastrointestinal Problems.
- Blood sugar changes.
- Hormone fluctuation which worsen menstrual cycles.

Depending on the type of autoimmune disease, an increase in the size of an organ or tissue or the destruction of an organ or tissue can result.

Diagnostic evaluation of immune dysfunctions

- Physical examination
- Symptoms of the individual
- Complete medical history.
- Laboratory tests:
- Specific antibody tests.
  - Thyroid function tests to test for thyroid disorders such as hyperthyroidism or hypothyroidism.
  - Rheumatoid factor test.
  - Acetylcholine receptor antibody test to test for myasthenia gravis.

- Blood tests, skin tests, bone marrow biopsy.

⌗ Specific lab test as:

★ **Leukocytes, lymphocytes test.**

★ **Humoral antibody, mediated immunity test.**

★ **HIV infection test:** The standard HIV test looks for antibodies in a person's blood. When HIV (which is a virus) enters a person's body, special proteins are produced. These are called antibodies. Antibodies are the body's response to an infection. So if a person has antibodies to HIV in their blood, it means they have been infected with HIV. The only exception might be an HIV negative baby born to a positive mother. Babies retain their mother's antibodies for up to 18 months, so may test positive on an HIV antibody test, even if they are actually HIV negative. This is why babies born to positive mothers may receive a PCR test after birth.

Most people develop detectable HIV antibodies within 6 to 12 weeks of infection. In very rare cases, it can take up to 6 months. It is exceedingly unlikely that someone would take longer than 6 months to develop antibodies.

ELISA:

The **enzyme-linked immunosorbent assay** (**ELISA**) is a test that uses antibodies and color change to identify a substance. The ELISA is a fundamental tool of clinical immunology, and is used as an initial screen for HIV detection. Based on the principle of antibody-antibody interaction, this test allows for easy visualization of results and can be completed without the additional concern of radioactive materials use.

Immunosuppressive therapy

- Definition
- Purpose
- Classification of immunosuppressive drugs
- Patient teaching and nursing management

Immunosuppressive therapy

Definition:

Immunosuppressive drugs, immunosuppressive agents, or immunosuppressants are drugs that inhibit or prevent activity of the immune system. They are commonly used to prevent production of antibodies (protein produced by immune system) in response to the presence of antigens.

Purpose:

- Prevent the rejection of transplanted organs and tissues (e.g., bone marrow, heart, kidney, liver)
- Treat autoimmune diseases or diseases that are most likely of autoimmune origin (e.g., rheumatoid arthritis, multiple sclerosis, myasthenia gravis, systemic lupus erythematosus).
- Treat some other non-autoimmune inflammatory diseases (e.g., long term allergic asthma control).

Classification of immunosuppressants

- **glucocorticoids as;(** Hydrocortisone, Dexamethasone , Prednisone**)**
- **calcineurin inhibitors (**drugs acting on immunophilins**)**
- As; Tacrolimus& Cyclosporine
- **cytostatics** As;**(**Azathioprine**,**Cyclophosphamide Methotrexate)
- **Sirolimus**
- **antibodies**
- **Other drugs (**opioids, interferons**)**

Patient teaching and nursing management :

1. Patients taking any of immunosuppressants should be encouraged to minimize the risk of infection , and encourage to give report of any fever, sore throat , chills , joint pain to doctors because this may indicate sever infection .
2. Patient should be told to take medication with meals or mixed with chocolate to prevent GI upset .
3. Patient who receive corticosteroid should be encourage to get adequate rest, exercise and nutrition.
4. Patient should be monitor frequently for weight gain, change in nutrition status , electrolyte imbalance .
5. Patient should be teach to avoid aspirin intake.
6. Inform the patient about expected beneficial and adverse reaction before starting immunosuppressive therapy
7. Take carefully history to determine the presence of allergies to immunosuppressive therapy .
8. Carefully check the patient for adverse reaction include inspection for gums hyperplasia , change in blood pressure level.
9. Monitor for elevated serum creatinine and BUN and inform the patient about the important of receiving periodic test of kidney function to determine the presence of nephrotoxicity .
10. Cyclosporine promote hair growth assure the patient that is effect is reversible.

11. Patient receive immunosuppressive need emotional support and assistance in accepting a chronic health education that require life long medication .
12. Patient should be inform about the important of maintaining the drug regimen .
13. Observe the patient for swelling of legs and shortness of breath which indicate cardio toxicity.
14. Monitor patient for neuromuscular status, watch for numbness, tingling, pain in finger and toes , headache , double vision and other early signs of neurotoxicity .
15. Immunosuppressant drugs lower a person's resistance to infection and can make infections harder to treat. The drugs can also increase the chance of uncontrolled bleeding.
16. The prescribing physician should be immediately informed if signs of infection, such as fever or chills, cough or hoarseness, pain in the lower back or side, or painful or difficult urination, bruising or bleeding, blood in the urine, bloody or black, tarry stools occur.
17. Other ways of preventing infection and injury include washing the hands frequently, avoiding any things which cause injuries, and being careful when using knives, razors, fingernail clippers or other sharp objects. Avoiding contact with people who have infections is also important.
18. Immunosuppressant drugs may cause the gums to become tender and swollen or to bleed. If this happens, a physician or dentist should be notified.
19. Regular brushing, flossing, cleaning and gum massage may help prevent this problem.
20. A dentist can provide advice on how to clean the teeth and mouth without causing injury.

Antihistamines

- Definition
- Pharmacological effects and uses
- Contraindications
- Patient teaching and nursing management

Antihistamines

Definition:

A drug that inhibits the action of histamines, reducing inflammation, swelling and other effects often experienced as an allergic reaction.

Pharmacological effects and uses:

- **first-generation antihistamines**

It respond to an acute attack or over-response by the immune system.Reduce capillary fragility that reduces redness, associated with allergic reactions).

Reduce histamine-induced secretions, including excessive tears and salivation and also, bind non-selectively to H1 receptors in the central nervous system as well as to peripheral receptors, and can produce sedation, inhibition of nausea and vomiting. May be used as a nighttime sedative, the relieve anxiety prior to surgery, as an antinauseant for control of drug induced Parkinsonism and in liquid form for control of coughs. As, phenothiazine.

- **second-generation antihistamines**

It bind only to peripheral H1 receptors, and reduce allergic response with little or no sedation, no central action, and are used only for treatment of allergic reactions. As, Loratidine (Claritin), Cetirizine (Zyrtec).

Side effects

Central nervous system: reactions include drowsiness, sedation, dizziness, faintness, disturbed coordination, lassitude, confusion, restlessness, excitation, tremor, seizures, headache, insomnia, euphoria, blurred vision, hallucinations, disorientation, disturbing dreams/nightmares, schizophrenic-like reactions, weakness, vertigo, hysteria, nerve pain, and convulsions. Overdoses may cause involuntary movements. Other problems have been reported.

Gastrointestinal problems: include dry mouth, increased appetite, decreased appetite, nausea, vomiting, diarrhea, and constipation.

Eye: corneal irritation due to decreases lacrimation.

Respiratory : chest tightness; wheezing; nasal stuffiness;, nose and throat; sore throat; respiratory depression; sneezing; and a burning sensation in the nose.

Hematologic reactions: are **rare**, but may be severe. These include anemia, or breakdown of red blood cells; reduced platelets; reduced white cells; and bone marrow failure.

Contraindications

- First-generation antihistamines possess anti-muscarinic activity and therefore must be used with caution in:
    - Benign prostatic hyperplasia and urinary retention

- Acute glaucoma
- Pyloric outflow obstruction
- Hepatic and/or renal impairment - dose reduction should be considered
- Epilepsy - use with caution
- Hepatic and renal impairment
- Pregnancy (teenager) and lactation antihistamines should be prescribed only where absolutely necessary in pregnant patients; avoid in first trimester.

child and caregiver teaching and nursing management :

- Teach teenager to avoid driving, or engaging in any hazardous activities until CNS response to drug treatment is stabilized.
- Suggest sucking on hard candy or ice chips for relief of dry mouth.
- Encourage use of sunscreen and hat while outdoors.
- Assess for urinary retention; monitor urinary output.
- Administer medication with food or milk to decrease GI distress.
- Increase fluid intake.
- Instruct patient to sit and relax a few minutes before activity if palpitation occur.
- Encourage use of humidification at home if nasal congestion occur.
- Instruct patient to rise from a sitting position slowly if hypotension occur.

Systemic Lupus Erythematosus (SLE)

It is a chronic inflammatory connective tissue disorder that can involve joints, kidneys, mucous membranes, and blood vessel walls.
Problems in the joints, nervous system, blood, skin, kidneys, gastrointestinal tract, and other tissues and organs can develop.
Lupus occurs in all parts of the world but may be more common among blacks and Asians.

Symptoms

Symptoms vary greatly from person to person. Symptoms may begin suddenly with fever, resembling a sudden, severe (acute) infection. Or symptoms may develop gradually over months or years with episodes

- Migraine-type headaches, epilepsy, or severe mental disorders (psychoses) may be the first abnormalities that are noticed.

Lupus symptoms are usually chronic and relapsing. The following are the most common symptoms of lupus. However, each child may experience symptoms differently. Symptoms may include:

- Malar rash - a rash shaped like a butterfly that is usually found on the bridge of the nose and the cheeks.
- Discoid rash - a raised rash found on the head, arms, chest, or back.
- Fever
- Inflammation of the joints
- Sunlight sensitivity
- Hair loss
- Mouth ulcers
- Fluid around the lungs, heart, or other organs
- Kidney problems
- Low white blood cell or low platelet count
- Raynaud's phenomenon - a condition in which the blood vessels of the fingers and toes go into spasm when triggered by factors such as cold, stress, or illness.
- Weight loss
- Nerve or brain dysfunction
- Anemia

How is lupus diagnosed? (Children's Hospital of Pittsburgh2011)

- Medical history
- Reported symptoms
- Physical examination that may include:

- Blood test - to detect for certain antibodies that are present in most people with lupus.
- blood and urine tests - to assess kidney function.
- complement test - to measure the level of complement, a group of proteins in the blood that help destroy foreign substances (low levels of complement in the blood are often associated with lupus).
- X-rays - a diagnostic test which uses invisible electromagnetic energy beams to produce images of internal tissues, bones, and organs onto film.

Further, the **American College of Rheumatology** created a set of criteria to assist physicians in making a diagnosis of lupus. The child must have four of the eleven specific criteria to be diagnosed with lupus. It is important to remember that having some of the following symptoms does not mean that your child has lupus. The criteria include:

- Malar rash - a rash shaped like a butterfly that is usually found on the bridge of the nose and the cheeks.
- Discoid rash - a raised rash usually found on the head, arms, chest, or back.
- Sunlight sensitivity.
- Mouth ulcers.

- Inflammation of the joints.
- Heart or lung involvement.
- Kidney problems.
- Seizures or other neurological problems.
- Positive blood tests.
- Changes in normal blood values.

PREVENTION

Because the cause of SLE is unknown, no one knows how to prevent it. Flares of lupus may be reduced by avoiding sun exposure (wearing strong sunscreen, hats, long-sleeved shirts in the sun), getting adequate sleep, and taking recommended medications. Risk of osteoporosis may be reduced by taking calcium and vitamin D.

Treatment for lupus: (Children's Hospital of Pittsburgh2011)

- There is no cure for lupus.

If lupus symptoms are mild, treatment may not be necessary, other than possibly nonsteroidal anti-inflammatory medications (NSAIDs) for joint pain. Other treatment may include:

- Corticosteroids (to control inflammation).
- Immunosuppressive medication (to suppress the body's autoimmune system).
- Liberal use of sunscreen, decreased time outdoors between 10:00 a.m. And 4:00 p.m., and wearing hats and long sleeves when outdoors, as about one-third of persons with lupus have the tendency to develop a rash in the sun.
- Rest, including at least eight to ten hours of sleep at night; naps and breaks during the day.
- Stress reduction.
- Well-balanced diet.
- Immediate treatment of infections.
- Hydroxychloroquine, quinacrine, chloroquine, or a combination of these medications.

Children with lupus should not receive immunizations with live viruses, including chickenpox, MMR (measles, mumps, rubella), and oral polio vaccines. Consult your child's physician regarding all vaccines.

Crohn's disease (CD)(Wong's 2013)

Crohn's disease is an inflammatory bowel disease. It is a chronic condition that may recur at various times over a lifetime. It usually involves the small intestine; most often the lower part called the ileum, inflammation may also affect the entire digestive tract, including the mouth, esophagus, stomach, duodenum, appendix or anus.

Crohn's disease is seen in children as young as seven years old. Males and females are affected equally. It appears to run in some families having a blood relative with some form of inflammatory bowel disease. In those who have a family history, it is very likely that Crohn's disease will begin in the teens and twenties.

The prevalence of Inflammatory bowel disease is between 12 years and 40 per 100,000 person with 25% of theses individuals beings diagnosed before 20 years of age .

children 6-17 years of age with CD appears to have more complicated disease course compared with that 0-5 years old children .

Etiology:

There are many theories regarding Crohn's disease, but none has yet been proven:

- Agent, perhaps a virus or bacteria, affects the body's immune system and triggers an inflammatory reaction in the intestinal wall.
- Abnormalities of the immune system, it is not known whether the immune problems are a cause or a result of the disease.
- Little proof that crohn's disease is caused by emotional distress or by an unhappy childhood.

Symptoms:

- Abdominal pain and cramping.
- Recurring diarrhea, not related to flu or other viral illness.
- Unexplained low grade fever.
- Anemia.
- Joint pain accompanied by redness.
- Eye inflammation, (if left untreated may lead to blindness).
- Abdominal swelling, accompanied by pain.
- Sudden severe constipation, with inability to pass gas.
- Fissures or openings on the skin with no known cause.
- Loss of appetite.
- Rectal bleeding.

Diagnosis

- **Blood tests** - to determine if there is anemia resulting from blood loss, or if there is an increased number of white blood cells, suggesting an inflammatory process.

- **Stool culture** - to determine if there is blood loss, or if an infection by a parasite or bacteria is causing the symptoms.
- **Endoscopy** to examine the inside of part of the digestive tract. Tissue samples from inside the digestive tract may also be taken for examination and testing.
- **Biopsy** - taking a sample of tissue (from the lining of the colon) for examination in a laboratory.
- **Upper gastrointestinal (GI) series** - examination of the esophagus, stomach and duodenum (the first section of the small intestine), often by drinking liquid barium.
- **Barium enema**, an x-ray of the abdomen shows strictures (narrowed areas), obstructions (blockages) and other problems.
- **Colonoscopy** - a test that uses a long, flexible tube with a light and camera lens at the end (colonoscopy) to examine inside the large intestine.

Treatment:

- **Medication.** Abdominal cramps and diarrhea may be helped by medications, which often lessen the inflammation in the colon. More serious cases may require steroid drugs, antibiotics or drugs that affect the body's immune system.
- **Diet and vitamin supplements.** No special diet has been proven effective for preventing or treating Crohn's disease. Some symptoms are made worse by milk , hot spices, or fiber, but this may not be true for every child . Large doses of vitamins are ineffective and may even cause harmful side effects.

  Children lose weight because of inadequate calorie intake, which can be due to several factors:
  - They may avoid eating to prevent pain associated with digestion.
  - They may absorb nutrients poorly through the inflamed digestive tract.
  - They have greater nutritional needs than average because of their disease.

  If favorite foods are eliminated from the diet, they may not feel enthusiastic about eating. Nutritional supplements or special high-calorie liquid formulas may sometimes be suggested, especially for children with delayed growth.
- **Feeding through a vein.** A small number of patients, who temporarily need extra nutrition, may need periods of feeding by vein (intravenously).
  - **Surgery.** Crohn's disease may be helped by surgery, but it cannot be cured by surgery. Surgery may help to either relieve chronic symptoms of active disease that does not respond to medical therapy or to correct complications, such as intestinal blockage, perforation, abscess or bleeding.

Nursing care:

1. If the patient is receiving parenteral nutrition, provide meticulous site care.
2. Give iron supplements and blood transfusion as ordered.
3. Administer medications as ordered.
4. Provide good patient hygiene and meticulous oral care if the patient is restricted to nothing by mouth.
5. Record fluid intake and output, weigh the patient daily.
6. If the patient is receiving TPN, monitor his condition closely.
7. Evaluate the effectiveness of medication administration.
8. Provide emotional support.
9. Emphasize the importance of adequate rest.
10. Give the patient a list of foods to avoid, including lactose-containing milk products, spicy or fried high-residue foods.
11. Teach the patient about the prescribed medications, their desires effects and possible adverse reactions.

Guidelines that may help control diarrhea include the following (Children Hospital Wisconsin 2012)

- Eat small, frequent meals.
- Avoid foods with milk or milk products containing lactose.
- Drink liquids at room temperature.
- Drink liquids between meals rather than with meals.
- Avoid caffeine.
- Avoid foods with sorbitol, xylitol and mannitol.
- Avoid concentrated sweets such as candy, cakes and pies.
- Avoid gas-producing foods .
- Include sources of soluble fiber such as:
    - Bananas.
    - Rice.

Practice parameters for the surgical management of Crohn's disease. National Guideline clearing house 2007

Perforation

- Patients with symptoms and/or signs of free perforation should undergo operation.
- Patients with large anteroparietal, interloop, intramesenteric, or retroperitoneal abscesses may be managed by antibiotics and percutaneous drainage.
- Patients with enteric fistulas and symptoms or signs of localized or systemic sepsis that persist despite appropriate medical therapy should be considered for operation. Asymptomatic patients with internal fistulas do not typically require surgery.

Obstruction

- Patients with symptomatic strictures in any location that do not appear amenable or responsive to medical therapy should undergo operation.
- Patients with asymptomatic strictures of the colon that cannot be adequately surveyed by biopsy and/or cytology brushing should be considered for operation.

Inflammation

- Patients with acute colitis and symptoms or signs of impending or actual perforation should undergo operation.
- Patients with acute colitis whose condition worsens despite appropriate medical therapy or fails to significantly improve after 48 to 96 hours of medical therapy should be considered for operation.

Hemorrhage

Patients with massive hemorrhage originating from any location may be managed by interventional radiologic and/or endoscopic techniques. If the patient is too unstable for this, or if this approach is unavailable or unsuccessful, the patient should undergo operation.

Neoplasia

1. Patients with long-standing Crohn's disease of the ileocolon or colon should undergo endoscopic surveillance. Patients with colitis often are advised to undergo a screening colonoscopy after eight to ten years of disease symptoms, and surveillance endoscopy every one to two years thereafter.
2. Patients with carcinoma, dysplasia-associated lesion or mass , high-grade dysplasia, or multifocal, low-grade dysplasia of the colon or rectum should undergo resection. The appropriate extent of the resection is unclear and could range from a limited segment that includes only the inflamed bowel to the entire colon and rectum.
3. Patients with long-standing Crohn's disease of the terminal ileum, ileocolon, or upper gastrointestinal locations should undergo biopsy of suspicious lesions at the time of strictureplasty.
4. **Growth Retardation and Extraintestinal Manifestations**
   - Prepubertal patients with significant growth retardation despite appropriate medical therapy should be considered for operation.
   - Patients with symptomatic disorders of the skin, mouth, eye, or joints who fail to respond to medical therapy should be considered for operation.
   - Patients with symptomatic, accessible strictures of the intestinal tract can be considered for endoscopic dilatation of the affected area**.**

Colon

1. Patients with disease of the colon that requires emergency or urgent surgery should typically undergo subtotal or total colectomy with end ileostomy

2. Patients with disease of the colon that requires elective surgery may undergo segmental or total colectomy with or without a primary anastomosis.
3. Patients who require surgery for disease of the rectum may undergo total proctocolectomy or proctectomy with creation of a stoma

References:

1. Gordon Sears W. Anatomy and physiology for nurses. London: Edward Arnold, 5th edition, 1974.
2. 2. Lewis Sharon. Medical- surgical Nursing. USA: Mosby, 2000.
3. Autoimmune Disease Avaliable at:
4. http://www.nativeremedies.com/ailment/autoimmune-disease-symptoms-info.html.
5. Systemic Lupus Erythematosus .Avaliable at: http://www.merckmanuals.com/home/bone_joint_and_muscle_disorders/autoimmune_disorders_of_connective_tissue/systemic_lupus_erythematosus_sle.html.
6. Autoimmune Disorders. Avaliable at: http://labtestsonline.org/understanding/conditions/autoimmune/.
7. Autoimmune diseases Avaliable at: http://www.childrenshospital.org/az/Site614/mainpageS614P0.html.
8. Systemic Lupus Erythematosus (SLE, or Lupus). Avaliable at: http://www.chp.edu/CHP/P01725.
9. Crohn's Disease. Avaliable at: http://www.chw.org/display/PPF/DocID/22816/router.asp.
10. Children with Crohn's Disease. Avaliable at: http://www.crohns.net/Miva/education/aboutcrohnskids.shtml.
11. Practice parameters for the surgical management of Crohn's disease. Avaliable at: http://guidelines.gov/content.aspx?id=12545.
12. Nursing Interventions for Crohn’s Disease. Avaliable at: http://nursingfile.com/nursing-care-plan/nursing-interventions/nursing-interventions-for-crohn%E2%80%99s-disease.html.