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QUESTIONS

1. Identify and briefly explain 5 primary immunodeficiency disorder.
2. Identify and briefly explain 2 secondary immunodeficiency disorder.

ANSWERS

1. Primary immunodeficiency disorder are as followed;
2. X-linked agammaglobulinemia (XLA)
3. Immunoglobulin A deficiency
4. Granulomatoses with polyangiitis (GPA)
5. Thymic hypoplasia
6. Severe combined immunodeficiency disease(SCID)
7. X-linked agammaglobulinemia (XLA); is a rare genetic disorder that affects the body's ability to fight [infection](https://en.wikipedia.org/wiki/Infection). It is much more common in males. In people with XLA, the [white blood cell formation process](https://en.wikipedia.org/wiki/Lymphopoiesis) does not generate mature [B cells](https://en.wikipedia.org/wiki/B_cell), which manifests as a complete or near-complete lack of proteins called [gamma globulins](https://en.wikipedia.org/wiki/Gamma_globulin), including [antibodies](https://en.wikipedia.org/wiki/Antibody), in their bloodstream. B cells are part of the [immune system](https://en.wikipedia.org/wiki/Immune_system) and normally manufacture antibodies (also called [immunoglobulins](https://en.wikipedia.org/wiki/Immunoglobulin)), which defend the body from infections by sustaining a [humoral immunity](https://en.wikipedia.org/wiki/Humoral_immunity" \o "Humoral immunity) response. Children are generally asymptomatic until 6–9 months of age when maternal IgG decreases. Patients with untreated XLA are prone to develop serious and even fatal infections. They can present with recurrent infections with Streptococcus pneumoniae, Haemophilus influenzae, Mycoplasma pneumoniae, hepatitis virus,  XLA is treated by infusion of human antibody.
8. **IMMUNOGLOBULIN A DEFICIENCY**; is a genetic [immunodeficiency](https://en.wikipedia.org/wiki/Immunodeficiency), a type of [hypogammaglobulinemia](https://en.wikipedia.org/wiki/Hypogammaglobulinemia). People with this deficiency lack [immunoglobulin A](https://en.wikipedia.org/wiki/Immunoglobulin_A) (IgA), a type of antibody that protects against infections of the mucous membranes lining the mouth, airways, and digestive tract. It is the most common of the primary antibody deficiencies. Most such persons remain healthy throughout their lives and are never diagnosed. Some patients with IgA deficiency have a tendency to develop recurrent sinopulmonary infections, gastrointestinal infections and disorders, allergies, and malignancies.
9. **GRANULOMATOSES WITH POLYANGIITIS (GPA)**; previously known as Wegener's granulomatosis (WG), is an extremely rare long-term systemic disorder that involves the formation of [granulomas](https://en.wikipedia.org/wiki/Granuloma) and [inflammation of blood vessels](https://en.wikipedia.org/wiki/Vasculitis) (vasculitis). It is a form of vasculitis that affects small- and medium-size vessels in many organs but most commonly affects the upper respiratory tract, lungs and kidneys. The signs and symptoms of GPA are highly varied and reflect which organs are supplied by the affected blood vessels. Typical signs and symptoms include [nosebleeds](https://en.wikipedia.org/wiki/Epistaxis), stuffy nose and crustiness of nasal secretions, and [inflammation of the uveal layer of the eye](https://en.wikipedia.org/wiki/Uveitis). Damage to the [heart](https://en.wikipedia.org/wiki/Heart), [lungs](https://en.wikipedia.org/wiki/Lung) and [kidneys](https://en.wikipedia.org/wiki/Kidney) can be fatal. GPA treatment depends on the severity of the disease. Severe disease is typically treated with a combination of [immunosuppressive medications](https://en.wikipedia.org/wiki/Immunosuppression) such as [rituximab](https://en.wikipedia.org/wiki/Rituximab) or [cyclophosphamide](https://en.wikipedia.org/wiki/Cyclophosphamide) and high-dose [corticosteroids](https://en.wikipedia.org/wiki/Corticosteroid) to control the [symptoms of the disease](https://en.wikipedia.org/wiki/Remission_(medicine)) and [azathioprine](https://en.wikipedia.org/wiki/Azathioprine), [methotrexate](https://en.wikipedia.org/wiki/Methotrexate), or rituximab to keep the disease under control. [Plasma exchange](https://en.wikipedia.org/wiki/Plasma_exchange) is also used in severe cases with damage to the lungs, kidneys, or intestines.
10. **THYMIC HYPOPLASIA**; Thymic hypoplasia is a condition where the [thymus](https://en.wikipedia.org/wiki/Thymus) is underdeveloped or involuted.  It often occurs as a sequel of another disorder, rather than as a standalone condition .
11. **SEVERE COMBINED IMMUNODEFICIENCY DISEASE**; Severe combined immunodeficiency (SCID) is a rare [genetic disorder](https://en.wikipedia.org/wiki/Genetic_disorder) characterized by the disturbed development of functional [T cells](https://en.wikipedia.org/wiki/T_cell) and [B cells](https://en.wikipedia.org/wiki/B_cell) caused by numerous genetic mutations that result in differing clinical presentations. SCID involves defective antibody response due to either direct involvement with [B lymphocytes](https://en.wikipedia.org/wiki/B_lymphocytes) or through improper B lymphocyte activation due to non-functional [T-helper cells](https://en.wikipedia.org/wiki/T-helper_cell). SCID is the most severe form of [primary immunodeficiencies](https://en.wikipedia.org/wiki/Primary_immunodeficiency) and there are now at least nine different known genes in which mutations lead to a form of SCID. It is also known as the bubble boy disease and bubble baby disease because its victims are extremely vulnerable to [infectious diseases](https://en.wikipedia.org/wiki/Infectious_diseases) and some have to live in a [sterile](https://en.wikipedia.org/wiki/Sterilization_(microbiology)) environment. SCID is the result of an immune system so highly compromised that it is considered almost absent. SCID patients are usually affected by severe bacterial, viral, or fungal infections early in life and often present with interstitial lung disease, chronic diarrhea, and failure to thrive. These babies, if untreated, usually die within one year due to severe, recurrent infections unless they have undergone successful [hematopoietic stem cell transplantation](https://en.wikipedia.org/wiki/Hematopoietic_stem_cell_transplantation). The most common treatment for SCID is [bone marrow transplantation](https://en.wikipedia.org/wiki/Bone_marrow_transplant), which has been very successful using either a matched related or unrelated donor, or a half-matched donor, who would be either parent. The half-matched type of transplant is called haploidentical. Haploidentical bone marrow transplants require the donor marrow to be depleted of all mature T cells to avoid the occurrence of [graft-versus-host disease](https://en.wikipedia.org/wiki/Graft-versus-host_disease) (GVHD).[[15]](https://en.wikipedia.org/wiki/Severe_combined_immunodeficiency#cite_note-titleTransplantation_immunology:_solid_organ_and_bone_marrow-15) Consequently, a functional immune system takes longer to develop in a patient who receives a haploidentical bone marrow transplant compared to a patient receiving a matched transplant.
12. The secondary immunodeficiency disorders are;
13. AIDS
14. Multiple myeloma
15. **AIDS**; AIDS refers to acquired immunodeficiency syndrome. AIDS is a disease that can develop in people with HIV. It’s the most advanced stage of HIV. But just because a person has HIV doesn’t mean they’ll develop AIDS. HIV kills CD4 cells. Healthy adults generally have a CD4 count of [500 to 1,500](https://www.hiv.va.gov/patient/diagnosis/labs-CD4-count.asp) per cubic millimeter. A person with HIV whose CD4 count falls below [200](https://www.hiv.va.gov/patient/diagnosis/labs-CD4-count.asp) per cubic millimeter will be diagnosed with AIDS. There’s no cure for AIDS, and without treatment, life expectancy after diagnosis is about [three years](http://www.cdc.gov/hiv/basics/whatishiv.html). This may be shorter if the person develops a severe opportunistic illness. However, treatment with antiretroviral drugs can prevent AIDS from developing. This may be shorter if the person develops a severe opportunistic illness. However, treatment with antiretroviral drugs can prevent AIDS from developing. If AIDS does develop, it means that the immune system is severely compromised. It’s weakened to the point where it can no longer fight off most diseases and infections. That makes the person vulnerable to a wide range of illnesses, such as [pneumonia](https://www.healthline.com/health/pneumonia), [tuberculosis](https://www.healthline.com/health/tuberculosis), [oral thrush](https://www.healthline.com/health/thrush), a fungal infection in the mouth or throat. Symptoms of AIDS can include recurrent fever, chronic fatigue, night sweats, chronic diarrhea etc.

b. **MULTIPLE MYELOMA**; Multiple myeloma is a type of cancer that affects plasma cells. Plasma cells are a type of white blood cell found in bone marrow, which is the soft tissue inside most of your bones that produces blood cells. In the bone marrow, plasma cells make antibodies. These are proteins that help your body fight off diseases and infections. Multiple myeloma occurs when an abnormal plasma cell develops in the bone marrow and reproduces itself very quickly. The rapid reproduction of malignant, or cancerous, myeloma cells eventually outweighs the production of healthy cells in the bone marrow. As a result, the cancerous cells begin to accumulate in the bone marrow, crowding out the healthy white blood cells and red blood cells. Like healthy blood cells, cancerous cells try to make antibodies. However, they can only produce abnormal antibodies called monoclonal proteins, or M proteins. When these harmful antibodies collect in the body, they can cause kidney damage and other serious problems. There are two main types of multiple myeloma and they’re categorized by their effect on the body:

An indolent myeloma causes no noticeable symptoms. It usually develops slowly and doesn’t cause bone tumors. Only small increases in M protein and M plasma cells are seen. And

A solitary plasmacytoma causes a tumor to form, typically in bone. It usually responds well to treatment, but needs close monitoring.

The symptoms of multiple myeloma vary depending on the person. Initially, symptoms may not be noticeable. However, as the disease progresses, most people will experience at least one of four major types of symptoms. These symptoms are generally referred to by the acronym CRAB, which stands for:

calcium

renal failure

anemia

bone damage

There’s no cure for multiple myeloma. However, there are treatments that can help ease the pain, reduce complications, and slow the progression of the disease. Treatments are only used if the disease is getting worse.