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Primary immunodeficiency disorder (PID) refers to a heterogeneous group of over 130 disorders that result from defects in immune system development and/or function.

Primary immunodeficiency disease is most often identified in infants and children, but it is possible that the condition is identified in adulthood. Primary immunodeficiency disease represents a diverse group of hundreds of diseases that can weaken the immune system. Most often, primary immunodeficiency disease results in increased susceptibility to both acute and chronic (long-term) infections.

Examples of primary immunodeficiency disorders

include:

- X-linked agammaglobulinemia (XLA)
- common variable immunodeficiency (CVID)
- severe combined immunodeficiency (SCID), which is known as alymphocytosis or “boy in a bubble” disease
- Selective IgA deficiency
- Chronic granulomatous disease.

1. **X-linked agammaglobulinemia** (**XLA**) is a condition that affects the immune system and occurs almost exclusively in males. People with **XLA** have very few B cells, which are specialized white blood cells that help protect the body against infection. Children with XLA are usually healthy for the first 1 or 2 months of life because they

are protected by antibodies acquired before birth from their mother. After this time, the maternal antibodies are cleared from the body, and the affected child begins to develop recurrent infections. In children with XLA, infections generally take longer to get better and then they come back again, even with antibiotic medications. The most common bacterial infections that occur in people with XLA are lung infections (pneumonia and bronchitis), ear infections (otitis), pink eye (conjunctivitis), and sinus infections (sinusitis).

2. **Common variable immune deficiency** (**CVID**) is a disorder that impairs the immune system. People with **CVID** are highly susceptible to infection from foreign

invaders such as bacteria, or more rarely, viruses and often develop recurrent infections, particularly in the lungs, sinuses, and ears. Pneumonia is common in people with CVID. Over time, recurrent infections can lead to chronic lung disease. Affected individuals may also experience infection or inflammation of the gastrointestinal tract, which can cause diarrhea and weight loss. Abnormal accumulation of immune cells causes enlarged lymph nodes (lymphadenopathy) or an enlarged spleen (splenomegaly) in some people with CVID. Immune cells can accumulate in other organs, forming small lumps called granulomas.

Approximately 25 percent of people with CVID have an

autoimmune disorder, which occurs when the immune system malfunctions and attacks the body's tissues and organs. The blood cells are most frequently affected by autoimmune attacks in CVID; the most commonly occurring autoimmune disorders are immune thrombocytopenia, which is an abnormal bleeding disorder caused by a decrease in cells involved in blood clotting called platelets, and autoimmune hemolytic anemia, which results in premature destruction of red blood cells.

3. **Severe Combined Immunodeficiency** (**SCID**) is an inherited primary **immunodeficiency** disease (PIDD) that typically presents in infancy results in profound **immune deficiency** condition resulting in a weak immune system that is unable to fight off even mild

infections. It is considered to be the most serious PIDD.

4. **Selective IgA deficiency:** Selective IgA deficiency is an immune system condition in which you lack or don't have enough immunoglobulin A (IgA), a protein that fights infection (antibody). Most people with selective IgA deficiency don't have recurrent infections. However, some people who have IgA deficiency experience pneumonia, ear infections, sinus infections, allergies, asthma and diarrhea.

Autoimmune diseases, in which your immune system attacks particular organs or tissues in your own body, can be found with selective IgA deficiency. Common autoimmune conditions found with IgA deficiency include rheumatoid

arthritis, lupus, celiac disease or inflammatory bowel disease.

5. **Chronic granulomatous disease:** Chronic Granulomatous Disease (CGD) is an inherited primary immunodeficiency disease (PIDD) which increases the body's susceptibility to infections caused by certain bacteria and fungi. Granulomas are masses of immune cells that form at sites of infection or inflammation. People with CGD are unable to fight off common germs and get very sick from infections that would be mild in healthy people. This is because the presence of CGD makes it difficult for cells called neutrophils to produce hydrogen peroxide. The immune system requires hydrogen peroxide to fight

specific kinds of bacteria and fungi.

Secondary immunodeficiency disorders happen when an outside source like a toxic chemical or infection attacks your body.
Examples of secondary immunodeficiency disorders include:

- AIDS
- cancers of the immune system, like leukemia
- immune-complex diseases, like viral hepatitis: **Immune complex diseases** encompass a diverse group of inflammatory conditions characterised by antigen–antibody deposition and attendant activation of complement. Common manifestations include glomerulonephritis, synovitis and dermal vasculitis. Many

patients present with constitutional symptoms and less specific signs.

2. **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.