Understanding PIDD

Primary Immunodeficiency Disease (PIDD)

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What is Primary Immunodeficiency?

Primary Immunodeficiency (PIDD or PID) is a disease that weakens a person's ability to fight off the germs that make us sick. PIDD occurs when certain parts of a person's immune system are missing or don't work like they should.

A person's immune system is responsible for defending the body from germs like bacteria and viruses that cause infections. This defense system is extremely complex, with many different types of cells and molecules that can fight off a wide variety of attacks.

When harmful germs enter our body, the immune system sends tiny molecules, called antibodies, through the blood to destroy them. These antibodies also learn how to tell the difference between the cells that our body makes naturally and the outside invaders that are bad for us.

In people with PIDD, the immune system may not make enough antibodies or may be missing some key components. Because of this, people with PIDD get sick more frequently than normal and may have trouble fighting off infections to get better.

Who is affected by PIDD?

PIDD is inherited (passed on from parents to children) and there are many different versions of the disease. Over 200 different forms have been identified. Some versions are severe and make it impossible for a person to fight off germs, while other versions can be very mild.

What are the symptoms of PIDD?

People with PIDD develop frequent infections, and these infections may be more severe and harder to cure than normal. People with PIDD may notice frequent pneumonia and bronchitis, ear and sinus infections, sore throats, coughs, colds and flu, fungal infections, rare viruses, and slowly healing wounds. Often, the same type of infection may come back again and again. Severe infections may require hospitalization.

Other symptoms include:

- An unusually high number of infections that require treatment
- Infections that do not respond to treatment, especially those requiring hospitalization
- Slow growth or lower-than-normal body weight in children
- A family history of frequent infections or PIDD
- Persistent thrush in the mouth or fungal infections on skin

How is PIDD diagnosed?

Very severe versions of PIDD are usually detected at birth with hospital testing, and patients are put on treatment immediately.

In more mild cases, it may be difficult to identify symptoms of PIDD. It is estimated that 500,000 people have PIDD in the US, and that half of those cases have not been diagnosed.

For people with PIDD, early diagnosis is the key to staying healthy and avoiding more severe consequences.

Common methods of diagnosis include:

- A review of the patient's medical history to see what type, frequency and severity of infections the person has had
- A physical exam. Children with PIDD may be underweight and grow more slowly than normal
- A review of the patient's family history to identify other family members who may have PIDD-like symptoms
- Blood tests to measure the level and make-up of the immune system
- Vaccines to test immune response

How is PIDD treated?

There is presently no cure for PIDD, but it can be effectively treated so that patients achieve a normal standard of living. PIDD treatments include:

- Oral or intravenous antibiotics to fight off active infections
- Enzyme replacement therapy
- Immunoglobulin (IVIg or IgG)

What is Immunoglobulin (IgG)?

Immunoglobulin (IgG) is made of purified antibodies, which are proteins made by the immune system to defend the body against infection. Since the immune systems in people with PIDD can't make enough of the molecules that fight infection, they take IgG to supplement their low or missing antibodies.

IVIg is IgG administered intravenously (through the vein), usually by a specially trained nurse. IgG can also be provided in a patient-administered format called Sub-Cutaneous Immunoglobulin (SCIg), in which a patient or parent gives injections of IgG. Both of these therapies can be done in the comfort of the patient's home.

For more information, please contact the Allergy and Immunology Awareness Program (AIAP) at:

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