About The Immune Deficiency Foundation of New Zealand (IDFNZ)

Our Promise

IDFNZ is a not-for-profit Organization dedicated to providing New Zealand families with Support, Advocacy and Education on Primary Immune Deficiency Disorders.

Patient membership: is free as we believe the price to be eligible to join is high enough.

Support: We have regional support staff who keep in direct contact with all members as required. They have direct experience of PI and can relate to the day-to-day issues facing the families, offering practical support, and understanding.

We are also able to match families with similar disorders if requested and organize various get togethers to bring groups of families together.

We issue a regular newsletter to all members and keep in touch to advise on any current issues affecting members.

The youth section of IDFNZ, is the **KIDS Foundation**, supporting PI children (0-18 yrs.) and their families.

Advocacy: When you live with chronic ill health, it often feels like you are battling alone to gain a decent quality of life. We have found that by combining our efforts we are successful in gaining what our families need. When you join **IDFNZ** we ask that you do not hesitate to use the advocacy service if there are problems accessing health care or gaining income support.

Education: It is known that information can make all the difference between coping with adversity and "going under" with the stress. **IDFNZ** is dedicated to the provision of latest information on disorders affecting PI members, therefore empowering families to make well informed life affecting decisions.

PI Awareness: Dedicated to raising awareness of PI symptoms, diagnosis, and treatment. IDFNZ offers regular information and seminars to medical professionals and the public.

IDFNZ has access to an expert medical advisory panel.

PI Research: IDFNZ supports PI research and testing in New Zealand.

IDFNZ is a member of **IPOPI**.

For more information for patients or medical professionals wishing to join IDFNZ:

Website: www.idfnz.org.nz

Email: info@idfnz.org.nz

Postal: PO Box 75076, Manurewa, Manukau 2243.

Facebook: https://www.facebook.com/IDFNZKidsFoundation/

IDFNZ is a fully registered charity CC 24570.

Want to support IDFNZ?

IDFNZ is not government funded and relies on the generosity of others through donations,grants and bequests. If you would like to support IDFNZ we welcome all expressions of interest from potential sponsors. All donations are fully tax deductible. Please email us to discuss at info@idfnz.org.nz

LIVING WITH PI

X-Linked Agammaglobulinemia (XLA)

IDFNZ
Immune Deficiencies Foundation of New Zealand

The Kids Foundation

X-Linked Agammaglobulinemia (XLA) also known as Bruton's Agammaglobulinemia or Congenital Agammaglobulinemia.

XLA is an inherited immunodeficiency disease in which patients lack the ability to produce antibodies, proteins that make up the gamma globulin or immunoglobulin fraction of blood plasma.

Antibodies are an integral part of the body's defense mechanism against bacteria, viruses etc. they are important in the recovery from infections Antibodies are produced by specialized cells in the body called plasma cells. The development of plasma cells proceeds in an orderly fashion from stem cells located in the bone marrow, to B- lymphocytes. On contact with a foreign substance called an antigen B- lymphocytes mature into the plasma cells that produce and secrete antibodies. Patients with XLA have mutations in a gene that is necessary for the normal development of B-lymphocytes.

Clinical presentation

As they lack antibodies, patients with X-Linked Agammaglobulinemia are prone to develop infections. Infections frequently occur at or near the surfaces of mucus membranes, such as the middle ear, sinuses and lungs, but can also involve the bloodstream or internal organs. They also may have recurrent gastrointestinal tract infections that can cause diarrhea. In patients without antibodies, any of these infections may also penetrate the mucosal surface, invade the bloodstream and spread to other organs deep within the body, such as the bones, joints or brain.

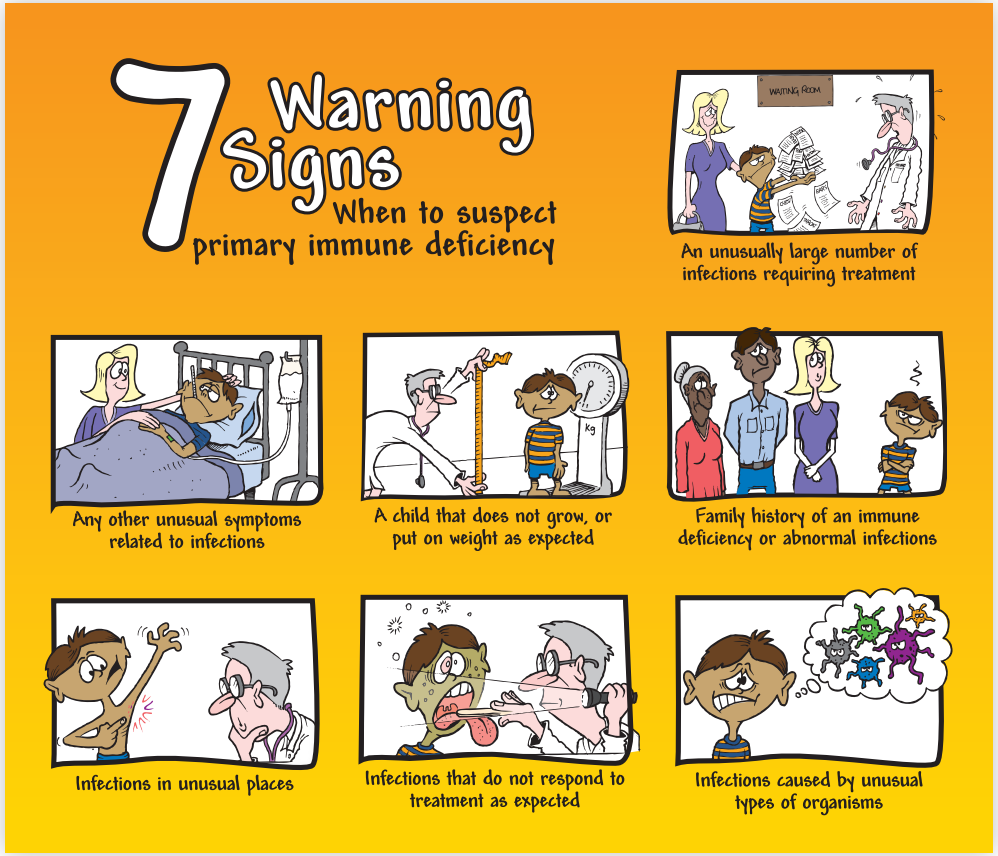
DIAGNOSIS When a patient is suspected of having XLA, the diagnosis is established by several tests. In XLA all the immunoglobulins (IgG, IgM, and IgA) are markedly reduced or absent in the blood. There is a complete absence of B-lymphocytes. This is the most reliable test, since it is not influenced by age, previous immunizations, or the IgG that the baby received across the placenta from the mother.

Inheritance

X- Linked Agammaglobulinemia is a genetic disease and as such can be inherited or passed on in a family. It is inherited as an X-linked recessive trait. This is why boys are affected and females are the carriers. Now that the precise gene that causes XLA has been identified, it is possible to test the female siblings of a patient with XLA, and other female relatives, to determine if they are carriers of the disease. Carriers of XLA usually have no symptoms but have a 50% chance of transmitting the disease to each of their sons.

Treatment

At this time the defective gene cannot be repaired or replaced. However, patients with XLA can be given some of the antibodies that they cannot make. The antibodies are supplied in the form of gamma globulins (or immunoglobulins) and can be given directly into the blood stream "intravenously" or just under the skin "subcutaneously". Gamma globulin is extracted from a large pool of human plasma consisting mostly of IgG and containing all the important antibodies present in the normal population.



Recurrent or chronic infections of the mucus membranes, such as sinusitis or pneumonia, can occur in some patients with XLA despite the use of gamma globulin. In these patients, it may be necessary to use long courses of antibiotics or other therapies.

Finally, patients with XLA should not receive any live viral vaccines, such as live polio, or the measles, mumps, rubella (MMR) vaccine. Although uncommon, it is possible that live vaccines in XLA patients can transmit the diseases that they were designed to prevent.

Expectations

Most X-Linked Agammaglobulinemia (XLA) patients who are receiving gamma globulin on a regular basis will be able to lead relatively normal lives. They do not need to be isolated or limited in their activities. Infections may require some extra attention from time to time, but children with XLA can participate in all regular school and extracurricular activities, and adults can have productive careers and families.

A full active lifestyle is to be encouraged and expected!