Research & Reviews in Pharmacy and Pharmaceutical Sciences e-ISSN: 2320-1215
p-ISSN: 2322-0112

Cause Diagnosis and Treatment of X-linked Agammaglobulinemia

Noah Richards*

Department of Pharmaceutical Science, Khalil Pharmacy, Jalalabad, Afghanistan

Commentary

Received: 25-Mar-2023, Manuscript No. JPPS-23-92808; **Editor assigned:** 27-Mar-2023, Pre QC No. JPPS-23-92808 (PQ); **Reviewed:** 10-Apr-2023, QC No. JPPS-23-92808; **Revised:** 24-May-2023, Manuscript No. JPPS-23-92808 (R); **Published:** 31-May-2023, DOI: 10.4172/2320-1215.12.2.002.

***For Correspondence** : Noah Richards, Department of Pharmaceutical Science, Khalil Pharmacy, Jalalabad, Afghanistan; **Email: richardsnoah@outlook.com**

Citation: Richards N. Cause Diagnosis and Treatment of X-linked Agammaglobulinemia. RRJ Pharm Pharm Sci. 2023;12:002.

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DESCRIPTION

Agammaglobulinemia is a group of inherited immunological disorders characterised by low antibody concentrations in the blood caused by a lack of specific lymphocytes in the blood and lymph. Antibodies are proteins (immunoglobulin's, IgM, IgG, and so on) that are essential components of the immune system. They are required for the immune system to do its function of defending the body against bacteria, viruses, and other foreign things. The specialised precursor cells that make gamma globulins fail to form or function effectively, resulting in a lack of mature lymphocyte cells known as B cells.

X-Linked Agammaglobulinemia (XLA), the considerably rarer X-linked agammaglobulinemia with growth hormone deficiency (approximately 10 instances recorded), and autosomal recessive agammaglobulinemia are the three kinds of Agammaglobulinemia (ARAG). All of these illnesses are characterised by a reduced immune system, which must be reinforced by gamma globulin injection in order to combat infections.

A mutation in the Bruton Tyrosine Kinase (BTK) gene, which is situated on the long arm of the X-chromosome, causes X-linked agammaglobulinemia. BTK is a member of the Tec family and encodes for signal transduction enzymes known as cytoplasmic non-receptor tyrosine kinases. BTK is required for the development of pre-B cells into mature B cells, which occurs in the bone marrow.

Treatment

There is no treatment for XLA. The purpose of treatment is to strengthen the immune system, prevent infections, and aggressively treat those that do develop.

Medications

Among the medications used to treat XLA are: Gammaglobulin. This is a type of protein found in blood that contains anti-infection antibodies. It is administered as an infusion into a vein every two to four weeks or as a weekly injection. Gammaglobulin reactions can cause headaches, chills, backaches, and nausea. During a viral infection, reactions are more likely.

Persons with XLA are given antibiotics on a regular basis to prevent infections. Others take antibiotics for bacterial infections for longer than people who do not have XLA. Because agammaglobulinemia patients are unable to manufacture specific antibodies, the primary medical treatment is immunoglobulin replacement. (Ig). Antibiotics used aggressively to treat bacterial infections may reduce long-term problems. Live virus vaccines (e.g., Measles, Mumps, and Rubella (MMR)) are not recommended for these patients and their families due to the risk of vaccine related illnesses. In patients with XLA, however, dendritic and T-cell responses to influenza are normal after administration of inactivated trivalent influenza vaccination. In several studies conducted throughout the world, Intravenous ig (IVIG) improves clinical status while decreasing the risk of dangerous infections such as pneumonia, meningitis, and GI illness. This appears to be the case with hypogammaglobulinemia caused by cancer. Most people with XLA who get immunoglobulin on a regular basis can live very normal lives.

Live virus immunisations, such as those for polio, measles, mumps, and rubella, should be avoided by people with XLA. Though uncommon, these vaccines have the potential to infect the recipient with the disease they were designed to avoid. This is true for the majority of B- and T-cell immunological deficiencies. Patients with XLA can handle viral infections in childhood due to intact T cell activity, although they are still more susceptible to specific enteroviruses, such as polio, coxsackie, and echo viruses. Chronic meningoencephalitis caused by these viruses can result in progressively progressing neurological deficits such as ataxia, paraesthesia, loss of cognitive skills, developmental regression, and sensorineural hearing loss. Other symptoms include fever, headaches, convulsions, and/or paralysis. Enteroviral infections of the muscles and skin can result in a dermatomyositis like condition with an erythematous rash and peripheral edoema.