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Opinion Article

Clinical Diagnosis and Symptoms Bruton's agammaglobulinemia

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DESCRIPTION

Bruton's agammaglobulinemia (XLA) is an inherited immunodeficiency disorder characterized by the absence of mature B cells, resulting in severe antibody deficiency and recurrent infections. It can appear in an infant as soon as the protective effect of maternal immunoglobulins off, which occurs between the ages of three and six months. X-linked agammaglobulinemia is a primary immunodeficiency disorder characterized by deficiencies in humoral immunity. It is caused by mutations in the Bruton Tyrosine Kinase (BTK) gene on the X chromosome. BTK is required for B-cell development and maturation; without it, maturation ceases before the B-cell stage, resulting in no mature B cells and, consequently, no antibodies. X-linked agammaglobulinemia, the first immunodeficiency disease discovered, is caused by a gene on the X chromosome that prevents the child from producing antibodies. Infections in the middle ear, sinuses, and lungs are common in children with this disease, but the bloodstream and internal organs may also be affected.

The child's immune system will be compromised as a result of this immunodeficiency disease, making it difficult for him or her to fight off bacterial and viral infections. This disease affects approximately one in every 10,000 children. Early detection and treatment may enable the child to lead a relatively normal and active life. Immunoglobulin deficiency results in absent antibody responses and increases the likelihood of developing bacterial infections. Encapsulated pyogenic bacteria are typically the culprits because antibodies opsonize them as a defense mechanism. *Streptococcus pneumoniae*, *Haemophilus influenzae* type B, *Streptococcus pyogenes*, and Pseudomonas species are among the most common pathogens.

The most common type of infection seen in these patients prior to diagnosis is recurrent otitis media. Certain enteroviruses are also protected from replication in the gastrointestinal tract and subsequent spread to the central nervous system by humoral immunity. As a result, there is an increased risk of developing enteroviral infections, which can manifest as meningoencephalitis, hepatitis, or dermatomyositis-like infections. However, as the use of intravenous immunoglobulin in these patients has increased, the incidence has decreased to a few isolated cases. Low (at least 2 standard deviations below the mean) levels of immunoglobulins (IgG, IgA, and IgM) and absent B cells (1% of all lymphocytes are $CD19^+$ cells, as detected by flow cytometry) are used to diagnose X-linked agammaglobulinemia. Transient neutropenia is also possible, and although genetic testing can help confirm a diagnosis, it is not required. For first-degree relatives, genetic testing is usually recommended. If the mutation has been identified in family members, a prenatal diagnosis can be obtained through mutational analysis of the chorionic villus, amniocentesis, or percutaneous umbilical cord blood samples. Children with X-linked agammaglobulinemia can develop severe infections and die at a young age. Children who develop chronic lung disease with bronchiectasis (airway widening and scarring) may live a shorter life. However, if diagnosed and treated early, the child should be able to live a relatively normal, active life without the need for isolation. The treatment will be determined by the child's symptoms, age, and overall health. It will also depend on the severity of the condition.

Antibodies are being replaced. This treatment provides your child with antibodies that they are unable to produce. It also aids in the prevention of the spread of infection. Any infections will be treated as soon as possible by the healthcare provider. Alternatively, child's health care provider may prescribe antibiotics before they develop an infection. There will be no live virus vaccines. This includes measles, mumps, rubella (MMR), rotavirus, smallpox, and chickenpox vaccines (Varicella). This is due to the possibility that your child will contract the disease against which the vaccine is designed to protect.

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