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Opinion Article

Glycine Encephalopathy and Nonketotic Hyperglycinemia

Nisan Ullar*

Department of Biotechnology, Mother Teresa Women's University, Kodaikanal, India

DESCRIPTION

Glycine encephalopathy is a genetic metabolic disorder characterized by unusually high amounts of the amino acid in glycine. In the brain, glycine acts as a chemical messenger to carry signals. Glycine encephalopathy is a rare autosomal recessive condition of glycine metabolism. The second most prevalent condition of amino acid metabolism, after the phenylketonuria, is also a glycine encephalopathy. Defects in the glycine cleavage system, and in the enzyme also involved in glycine catabolism, are the root cause of the illness. There are various illness variants with different symptom intensity and onset times.

Clinically, this illness is characterized by excessively high quantities of the amino acid glycine in physiological fluids and tissues, particularly the cerebrospinal fluid. The symptoms are solely neurological in origin. *AMT*, *GLDC*, or *GCSH* gene variations (mutations) that lead to a lack of the enzyme needed to break down glycine are the root cause of glycine encephalopathy. The diagnosis is made based on the symptoms, the elevated glycine levels, the lack of an enzyme, and genetic testing. It is autosomal recessive in nature. Treatment options include a ketogenic diet, N-methyl D-aspartate (*NMDA*) receptor site antagonists, sodium benzoate to lower glycine levels, and anti-seizure medications. The survivors of the classic type may suffer motor delays, very small heads, convulsions, and rigidity. About half of the infants with the classic form die during the first few weeks of life. In the transitory type symptoms may improve with time.

The biochemical characteristics of glycine encephalopathy, and to distinguish it from effects that occur in "Ketotic Hyperglycemia," the condition is frequently referred to as "Nonketotic Hyperglycinemia" (NKH) (seen in propionic acidemia and several other inherited metabolic disorders). The term "glycine encephalopathy" is frequently used to communicate efficiently because it better appropriately defines the disorder's clinical symptoms. Glycine builds up in human tissues and fluids due to a deficiency in the enzyme system that breaks down the amino acid glycine in Non-Ketotic Hyperglycinemia (NKH), a rare genetic metabolic condition. Both the complementary and progressive forms of NKH exist. Glycine encephalopathy has been identified in mild and transitory forms. After a viable period and neonatal period, mild forms appear in infancy or early childhood. Clinical features include seizures (in most cases) and relatively mild developmental delay. Although it has only sometimes been identified, transient glycine encephalopathy exhibits the same initial clinical and biochemical characteristics as the classic type. The majority of patients exhibit normal development during the transitory form, where increased CSF and plasma glycine levels partially or totally normalize.

Nonketotic Hyperglycinemia is a condition known as characterized by excessively high amounts of the chemical glycine in the body (hyperglycinemia). Glycine accumulates in excess in tissues and organs, especially the brain. Serious neurological issues attack those who are affected. There are two types of Nonketotic Hyperglycinemia: the severe form and the attenuated form. Although signs and symptoms might sometimes start in the first few months of life, both forms often start soon after birth. Only the attenuated form delays the onset of childhood. The severity of the symptoms and indications distinguishes the forms. More people experience severe Nonketotic Hyperglycinemia. Babies who are affected have significant lethargy, which becomes worse with time and can result in coma.

In the initial few days or weeks of life, they may also experience life-threatening respiratory issues and hypotonic muscular tone. When these early warning signs and symptoms are survived, the majority of children experience eating issues, abnormal muscle stiffness (spasticity), substantial intellectual disabilities, and difficult-to-control seizures. Children are most impacted. When these early warning signs and symptoms are survived, the majority of children experience nutrition difficulties, abnormal muscle stiffness, substantial intellectual disabilities, and difficult-to-control seizures. The majority of affected kids don't reach typical developmental milestones like sipping from a bottle, sitting up, or gripping things and any abilities they do learn could be lost over time.

Correspondence to: Nisan Ullar, Department of Biotechnology, Mother Teresa Women's University, Kodaikanal, India, E-mail: ulllar708@yahoo.com

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