

Hydroxyglutaric **acid** is a collective term that refers to a group of organic acids that share a hydroxyglutarate chemical structure. There are several forms of hydroxyglutaric acid, including:

Alpha-hydroxyglutaric acid (α -Hydroxyglutaric acid or α -HGA): It exists in two forms, D-alpha-hydroxyglutaric acid (D- α -HGA) and L-alpha-hydroxyglutaric acid (L- α -HGA). Elevated levels of D- α -HGA or L- α -HGA are associated with rare metabolic disorders known as D- α -hydroxyglutaric aciduria and L- α -hydroxyglutaric aciduria, respectively.

Beta-hydroxyglutaric acid (β -Hydroxyglutaric acid or β -HGA): It also exists in two forms, D-beta-hydroxyglutaric acid (D- β -HGA) and L-beta-hydroxyglutaric acid (L- β -HGA). Elevated levels of D- β -HGA or L- β -HGA are associated with rare metabolic disorders called D- β -hydroxyglutaric aciduria and L- β -hydroxyglutaric aciduria, respectively.

These hydroxyglutaric acids are organic acids derived from the metabolism of certain amino acids and organic compounds in the body. Accumulation of these acids can disrupt normal cellular processes and contribute to the development of various health conditions, including neurological symptoms and developmental delays.

The diagnosis of hydroxyglutaric acidurias is typically confirmed by measuring the levels of the specific hydroxyglutaric acid in urine, blood, or cerebrospinal fluid. Treatment focuses on managing symptoms and may include dietary modifications and supportive therapies.

It's important to note that elevated levels of hydroxyglutaric acid can also be seen in some types of brain tumors, such as gliomas, as a result of mutations in isocitrate dehydrogenase (IDH) enzymes. These tumors are characterized by the accumulation of 2-hydroxyglutarate (2-HG), a derivative of alpha-hydroxyglutaric acid, which has been implicated in tumor development and progression.

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