D-2-hydroxyglutarate dehydrogenase

alpha-Hydroxyglutaric acid

D-2-hydroxyglutarate Dehydrogenase is an enzyme involved in the metabolism of D-2-hydroxyglutarate (D-2-HG). This enzyme, also known as D2HGDH, catalyzes the conversion of D-2-HG to alpha-ketoglutarate (α-KG) in a process known as oxidation.

The primary function of D-2-hydroxyglutarate dehydrogenase is to regulate the levels of D-2-HG in cells. D-2-HG is an oncometabolite, meaning its accumulation can contribute to the development and progression of certain cancers. D2HGDH plays a role in mitigating the effects of elevated D-2-HG by converting it back to α-KG, which is a crucial metabolite involved in various cellular processes.

Mutations in the D2HGDH gene can impair the activity or expression of D-2-hydroxyglutarate dehydrogenase, resulting in decreased enzymatic function and reduced ability to metabolize D-2-HG. This can lead to the accumulation of D-2-HG in cells and contribute to the pathogenesis of diseases, such as D-2-hydroxyglutaric aciduria (D2HGA), a rare metabolic disorder characterized by high levels of D-2-HG in the body.

Research on D-2-hydroxyglutarate dehydrogenase and its role in cellular metabolism and disease is ongoing. Understanding the regulation and function of this enzyme can provide insights into the mechanisms underlying D-2-HG-associated disorders and potentially lead to the development of targeted therapies.