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Mitochondrial Hydroxyacyl-Coenzyme A Dehydrogenase Hyperinsulinism

National Cancer Institute

Source

National Cancer Institute. *Mitochondrial Hydroxyacyl-Coenzyme A Dehydrogenase Hyperinsulinism*. NCI Thesaurus. Code C131837.

Hyperinsulinism due to inactivating mutation(s) in the gene HADH, encoding mitochondrial (short-chain) hydroxyacyl-coenzyme A dehydrogenase, resulting in loss of inhibition of glutamate dehydrogenase (GDH). This condition is also characterized by protein-induced hypoglycemia, but in contrast to GLUD1-associated hyperinsulinism, hyperammonemia is absent.