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KD-Validated Anti-HADHA Mouse Monoclonal Antibody

Mouse monoclonal antibody
Catalog # AGI1958

Product Information

Application	WB, FC
Primary Accession	P40939
Reactivity	Rat, Human, Mouse
Clonality	Monoclonal
Isotype	Mouse IgG1
Clone Names	24GB6655
Calculated MW	83000
Gene Name	HADHA
Aliases	HADHA; Hydroxyacyl-CoA Dehydrogenase Trifunctional Multienzyme Complex Subunit Alpha; LCHAD; LCEH; MTPA; GBP; Hydroxyacyl-Coenzyme A Dehydrogenase/3-Ketoacyl-Coenzyme A Thiolase/Enoyl-Coenzyme A HydRatase (Trifunctional Protein), Alpha Subunit; Hydroxyacyl-CoA Dehydrogenase/3-Ketoacyl-CoA Thiolase/Enoyl-CoA HydRatase (Trifunctional Protein), Alpha Subunit; Mitochondrial Trifunctional Protein, Alpha Subunit; Trifunctional Enzyme Subunit Alpha, Mitochondrial; Long-Chain-3-Hydroxyacyl-CoA Dehydrogenase; Monolysocardiolipin Acyltransferase; Long-Chain 2-Enoyl-CoA HydRatase; 78 KDa Gastrin-Binding Protein; Gastrin-Binding Protein; HADH; Mitochondrial Long-Chain L-3-Hydroxyacyl-Coenzyme A (CoA) Dehydrogenase, Alpha Subunit; Mitochondrial Long-Chain 2-Enoyl-Coenzyme A (CoA) HydRatase, Alpha Subunit; 3-Ketoacyl-Coenzyme A (CoA) Thiolase, Alpha Subunit; Mitochondrial Trifunctional Enzyme, Alpha Subunit; 3-Oxoacyl-CoA Thiolase; EC 2.3.1.- ; TP-ALPHA; TP-Alpha; ECHA
Immunogen	Recombinant protein of human HADHA

Additional Information

Gene ID	3030
Other Names	Trifunctional enzyme subunit alpha, mitochondrial, 78 kDa gastrin-binding protein, Monolysocardiolipin acyltransferase, MLCL AT, 2.3.1.-, TP-alpha, Long-chain enoyl-CoA hydratase, 4.2.1.17, Long chain 3-hydroxyacyl-CoA dehydrogenase, 1.1.1.211, HADHA, HADH

Protein Information

Name	HADHA
Synonyms	HADH
Function	Mitochondrial trifunctional enzyme catalyzes the last three of the four reactions of the mitochondrial beta-oxidation pathway (PubMed:1550553,

PubMed:29915090, PubMed:30850536, PubMed:8135828, PubMed:31604922). The mitochondrial beta-oxidation pathway is the major energy-producing process in tissues and is performed through cycles of four consecutive reactions (PubMed:29915090). Each beta-oxidation cycle shortens the fatty acyl-CoA by two carbons, yielding one acetyl-CoA (for the citric acid cycle), one FADH(2), and one NADH (which donate electrons to the respiratory chain for ATP production) (PubMed:29915090). These cycles repeat until the chain is fully degraded to acetyl-CoA units (PubMed:29915090). Among the enzymes involved in this pathway, the trifunctional protein--responsible for the hydration, dehydrogenation, and thiolysis steps, shows specificity for long-chain fatty acids, such as those from dietary and stored fats (PubMed:30850536, PubMed:31604922). Mitochondrial trifunctional enzyme is a heterotetrameric complex composed of two proteins, the trifunctional enzyme subunit alpha/HADHA described here carries the 2,3-enoyl-CoA hydratase and the 3-hydroxyacyl-CoA dehydrogenase activities while the trifunctional enzyme subunit beta/HADHB bears the 3-ketoacyl-CoA thiolase activity (Probable) (PubMed:29915090, PubMed:30850536, PubMed:8135828). These activities have been experimentally confirmed on a few substrates derived from beta- oxidation of long-chain saturated fatty acids such as palmitate (hexadecanoate) and laurate (dodecanoate) (PubMed:1550553, PubMed:8135828, PubMed:8163672, PubMed:8651282). In addition, based on its established catalytic mechanism, and combined genetic interaction or mutant phenotype evidence, it is predicted to act also on other substrates, including long-chain unsaturated fatty acids such as oleate (9Z-octadecenoate), linoleate (9Z,12Z-octadecadienoate), linolenate (9Z,12Z,15Z-octadecatrienoate), and others (Probable) (PubMed:26474213). Independently of subunit beta, HADHA also exhibits a cardiolipin acyltransferase activity that participates in cardiolipin remodeling; cardiolipin is a major mitochondrial membrane phospholipid (PubMed:23152787, PubMed:31604922). HADHA may act downstream of Tafazzin/TAZ, that remodels monolysocardiolipin (MLCL) to a cardiolipin intermediate, and then HADHA may continue to remodel this species into mature tetralinoleoyl-cardiolipin (PubMed:31604922). Has also been proposed to act directly on MLCL; capable of acylating MLCL using different acyl-CoA substrates, with highest activity for oleoyl-CoA (PubMed:23152787).

Cellular Location

Mitochondrion. Mitochondrion inner membrane Note=Protein stability and association with mitochondrion inner membrane do not require HADHB.

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