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HADHA Rabbit mAb

Catalog # AP75525

Product Information

Primary Accession	P40939
Host	Rabbit
Clonality	Monoclonal Antibody
Calculated MW	83000

Additional Information

Gene ID	3030
Other Names	HADHA
Format	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.

Protein Information

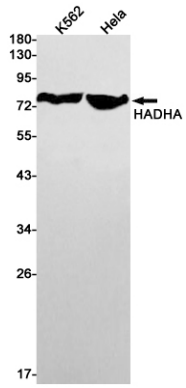
Name	HADHA
Synonyms	HADH
Function	Mitochondrial trifunctional enzyme catalyzes the last three of the four reactions of the mitochondrial beta-oxidation pathway (PubMed:1550553, PubMed:29915090, PubMed:30850536, PubMed:8135828, PubMed:31604922). The mitochondrial beta-oxidation pathway is the major energy-producing 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

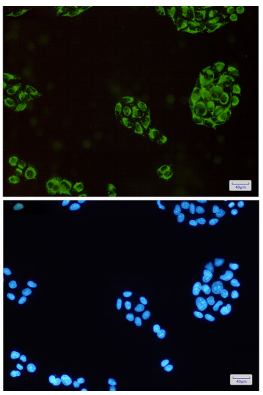
palmitate (hexadecanoate) and laurate (dodecanoate) (PubMed:1550553, PubMed:8135828, PubMed:8163672, PubMed:8651282). In addition, based on its established catalytic mechanism, and combined genetic interaction or mutant phenotype evidence, it is predicted to act also on other substrates, including long-chain 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.

Images





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