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HADHB Rabbit pAb

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Catalog # AP58270

Product Information

Application	WB
Primary Accession	P55084
Reactivity	Human, Mouse
Predicted	Rat, Pig, Dog, Rabbit, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	51294
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human HADHB
Epitope Specificity	231-330/474
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Mitochondrion. Mitochondrion inner membrane. Mitochondrion outer membrane. Endoplasmic reticulum.
DISEASE	Defects in HADHB are a cause of trifunctional protein deficiency (TFP deficiency) [MIM:609015]. The clinical manifestations are very variable and include hypoglycemia, cardiomyopathy and sudden death. Phenotypes with mainly hepatic and neuromyopathic involvement can also be distinguished. Biochemically, TFP deficiency is defined by the loss of all three enzyme activities of the TFP complex.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The HADHB gene encodes the beta subunit of the mitochondrial trifunctional protein, which catalyzes the last three steps of mitochondrial beta-oxidation of long chain fatty acids. The mitochondrial membrane-bound heterocomplex is composed of four alpha and four beta subunits, with the beta subunit catalyzing the 3-ketoacyl-CoA thiolase activity. Mutations in this gene result in trifunctional protein deficiency. The encoded protein can also bind RNA and decreases the stability of some mRNAs. The genes of the alpha and beta subunits of the mitochondrial trifunctional protein are located adjacent to each other in the human genome in a head-to-head orientation. Alternatively spliced transcript variants have been found; however, their full-length nature is not known.

Additional Information

Gene ID	3032
Other Names	Trifunctional enzyme subunit beta, mitochondrial, TP-beta, 3-ketoacyl-CoA thiolase, 2.3.1.155, 2.3.1.16, Acetyl-CoA acyltransferase, Beta-ketothiolase,

	HADHB
Dilution	WB=1:500-2000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

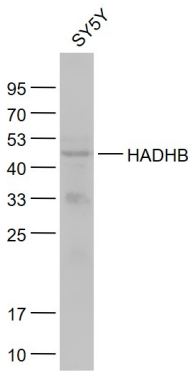
Name	HADHB
Function	Mitochondrial trifunctional enzyme catalyzes the last three of the four reactions of the mitochondrial beta-oxidation pathway (PubMed:1550553, PubMed:16423905, PubMed:26474213, PubMed:29915090, PubMed:30850536, PubMed:8135828). The mitochondrial beta-oxidation pathway is the major energy-producing process in tissues and is performed through cycles of four consecutive reactions (PubMed:26474213, 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:26474213, PubMed:29915090). These cycles repeat until the chain is fully degraded to acetyl-CoA units (PubMed:26474213, 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). Mitochondrial trifunctional enzyme is a heterotetrameric complex composed of two proteins, the trifunctional enzyme subunit alpha/HADHA carries the 2,3-enoyl-CoA hydratase and the 3-hydroxyacyl-CoA dehydrogenase activities while the trifunctional enzyme subunit beta/HADHB described here bears the 3-ketoacyl-CoA thiolase activity (PubMed:16423905, PubMed:26474213, PubMed:29915090, PubMed:30850536, PubMed:38372965, 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).
Cellular Location	Mitochondrion. Mitochondrion inner membrane Mitochondrion outer membrane. Endoplasmic reticulum. Note=Protein stability and association with membranes require HADHA

Background

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Images

SY5Y(Human) Cell Lysate at 30 ug



Primary: Anti-HADHB (AP58270) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 47 kD
Observed band size: 47 kD

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.