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HADHB Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP58270

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	P55084
Reactivity	Rat, Pig, Dog, Bovine
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.
SIMILARITY	Belongs to the thiolase family.
SUBUNIT	Octamer of 4 alpha (HADHA) and 4 beta (HADHB) subunits. Interacts with RSAD2/viperin.
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

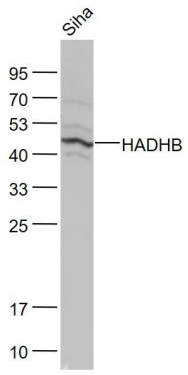
Gene ID	3032

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,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
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

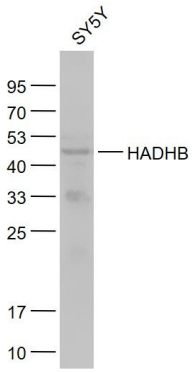
Name	HADHB
Function	Mitochondrial trifunctional enzyme catalyzes the last three of the four reactions of the mitochondrial beta-oxidation pathway (PubMed:29915090, PubMed:30850536, PubMed:8135828). The mitochondrial beta-oxidation pathway is the major energy-producing process in tissues and is performed through four consecutive reactions breaking down fatty acids into acetyl-CoA (PubMed:29915090). Among the enzymes involved in this pathway, the trifunctional enzyme exhibits specificity for long- chain fatty acids (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:29915090, PubMed:30850536, PubMed:8135828).
Cellular Location	Mitochondrion. Mitochondrion inner membrane Mitochondrion outer membrane. Endoplasmic reticulum. Note=Protein stability and association with membranes require HADHA

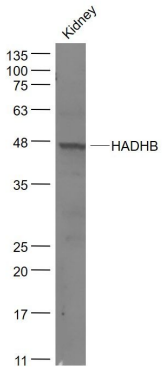
Images



Siha(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

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





Sample:
Kidney (Mouse) Lysate at 40 ug
Primary: Anti- HADHB (AP58270) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 47kD
Observed band size: 47 kD