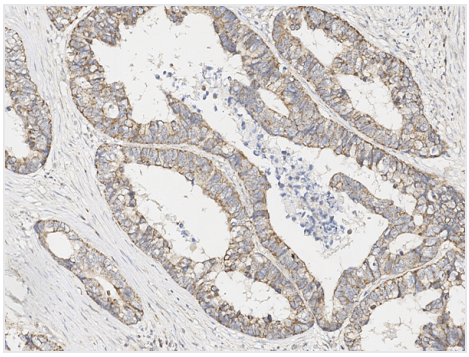
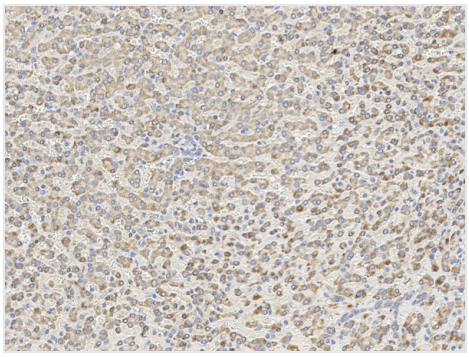
HADH Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

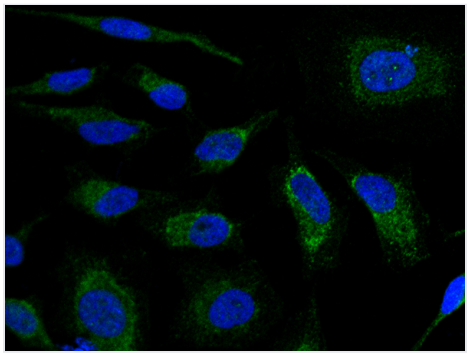
Product Name	HADH, Polyclonal Antibody
Full Product Name	HADH Antibody, Rabbit PAb, Antigen Affinity Purified
Product Gene Name	anti-HADH antibody
Product Synonym Names	Anti-HAD Antibody; Anti-HADH1 Antibody; Anti-HADHSC Antibody; Anti-HCDH Antibody; Anti-HHF4 Antibody; Anti-MSCHAD Antibody; Anti-SCHAD Antibody
Clonality	Polyclonal
Host	Rabbit
Species Reactivity	Human
Specificity	Human HADH
Preparation and Storage	This antibody can be stored at 2-8 degree C for one month without detectable loss of activity. Antibody products are stable for twelve months from date of receipt when stored at -20 to -80 degree C. Preservative-Free. Avoid repeated freeze-thaw cycles.
Molecular Weight	34,294 Da
OMIM	231530
3D Structure	ModBase 3D Structure for Q16836
Preparation	Produced in rabbits immunized with a synthetic peptide corresponding to the C-terminus of the Human HADH, and purified by antigen affinity chromatography.
Applications Tested/Suitable for anti-HADH antibody	Immunohistochemistry-Paraffin (IHC-P), Immunocytochemistry (ICC), Immunofluorescence (IF)
Related Product Information for anti-HADH antibody	HADH (Hydroxyacyl-CoA Dehydrogenase, also known as HADHSC) is a Protein Coding gene. 2 alternatively spliced human isoforms have been reported. This gene is a member of the 3-hydroxyacyl-CoA dehydrogenase gene family. The encoded protein functions in the mitochondrial matrix to catalyze the oxidation of straight-chain 3-hydroxyacyl-CoAs as part of the beta-oxidation pathway. HADH is a key enzyme in fatty acid oxidation. It is widely expressed in fat, kidney, and other tissues. The downregulation of HADH promotes gastric cancer progression via activation of the Akt signaling pathway. Recessive mutations in the hydroxyacyl-CoA dehydrogenase (HADH) gene encoding the enzyme 3-hydroxyacyl-CoA dehydrogenase are a rare cause of diazoxide-responsive hyperinsulinemic hypoglycemia (HH).
Other Notes	Small volumes of anti-HADH antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for HADH purchase	MBS8124304 is an antibody targeting HADH. Also known as HADH Antibody, Rabbit PAb, Antigen Affinity Purified. Reactive in Human. Host: Rabbit.



Immunohistochemistry (IHC)-Paraffin of anti-HADH antibody
Immunochemical staining of human HADH in human carcinoma of sigmoid with rabbit polyclonal antibody at 1:100 dilution, formalin-fixed paraffin embedded sections.



Immunohistochemistry (IHC)-Paraffin of anti-HADH antibody
Immunochemical staining of human HADH in human liver with rabbit polyclonal antibody at 1:100 dilution, formalin-fixed paraffin embedded sections.



Immunofluorescence (IF) of anti-HADH antibody
Immunofluorescence staining of HADH in Hela cells. Cells were fixed with 4% PFA, permeabilzed with 0.3% Triton X-100 in PBS,blocked with 10% serum, and incubated with rabbit anti-Human HADH polyclonal antibody (dilution ratio 1:100) at 4 degree C overnight. Then cells were stained with the Alexa Fluor488-conjugated Goat Anti-rabbit IgG secondary antibody (green) and counterstained with DAPI (blue).Positive staining was localized to Cytoplasm.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for HADH. It may not necessarily be applicable to this product.

NCBI GI #	296179429
NCBI GeneID	3033
NCBI Accession #	NP_005318.3
UniProt Primary Accession #	Q16836
UniProt Secondary Accession #	O00324; O00397; O00753; Q4W5B4; J3KQ17
UniProt Related Accession #	Q16836
GenBank Nucleotide #	NM_005327.4
NCBI Official Full Name	hydroxyacyl-coenzyme A dehydrogenase, mitochondrial isoform 2
NCBI Official Synonym Full Names	hydroxyacyl-CoA dehydrogenase
NCBI Official Symbol	HADH
NCBI Official Synonym Symbols	HAD; HCDH; HHF4; HADH1; SCHAD; HADHSC; MSCHAD
UniProt Protein Name	Hydroxyacyl-coenzyme A dehydrogenase, mitochondrial
UniProt Synonym Protein Names	Medium and short-chain L-3-hydroxyacyl-coenzyme A dehydrogenase; Short-chain 3-hydroxyacyl-CoA dehydrogenase
Protein Family	Hydroxyacyl-coenzyme A dehydrogenase

UniProt Gene Name	HADH
UniProt Synonym Gene Names	HAD; HADHSC; SCHAD; HCDH
UniProt Entry Name	HCDH_HUMAN
NCBI Protein Information	hydroxyacyl-coenzyme A dehydrogenase, mitochondrial; short-chain 3-hydroxyacyl-CoA dehydrogenase; L-3-hydroxyacyl-Coenzyme A dehydrogenase, short chain; medium and short-chain L-3-hydroxyacyl-coenzyme A dehydrogenase
NCBI Summary for HADH	This gene is a member of the 3-hydroxyacyl-CoA dehydrogenase gene family. The encoded protein functions in the mitochondrial matrix to catalyze the oxidation of straight-chain 3-hydroxyacyl-CoAs as part of the beta-oxidation pathway. Its enzymatic activity is highest with medium-chain-length fatty acids. Mutations in this gene cause one form of familial hyperinsulinemic hypoglycemia. The human genome contains a related pseudogene of this gene on chromosome 15. [provided by RefSeq, May 2010]
UniProt Comments for HADH	HADHSC: Plays an essential role in the mitochondrial beta- oxidation of short chain fatty acids. Exerts it highest activity toward 3-hydroxybutyryl-CoA. Defects in HADH are the cause of 3-alpha-hydroxyacyl-CoA dehydrogenase deficiency (HADH deficiency). HADH deficiency is a metabolic disorder with various clinical presentations including hypoglycemia, hepatoencephalopathy, myopathy or cardiomyopathy, and in some cases sudden death. Defects in HADH are the cause of familial hyperinsulinemic hypoglycemia type 4 (HHF4); also known as persistent hyperinsulinemic hypoglycemia of infancy (PHHI) or congenital hyperinsulinism. HHF is the most common cause of persistent hypoglycemia in infancy and is due to defective negative feedback regulation of insulin secretion by low glucose levels. It causes nesidioblastosis, a diffuse abnormality of the pancreas in which there is extensive, often disorganized formation of new islets. Unless early and aggressive intervention is undertaken, brain damage from recurrent episodes of hypoglycemia may occur. HHF4 should be easily recognizable by analysis of acylcarnitine species and that this disorder responds well to treatment with diazoxide. It provides the first 'experiment of nature' that links impaired fatty acid oxidation to hyperinsulinism and that provides support for the concept that a lipid signaling pathway is implicated in the control of insulin secretion. Belongs to the 3-hydroxyacyl-CoA dehydrogenase family. 2 isoforms of the human protein are produced by alternative splicing. Protein type: Amino Acid Metabolism - lysine degradation; Lipid Metabolism - fatty acid elongation in mitochondria; Carbohydrate Metabolism - butanoate; Oxidoreductase; Lipid Metabolism - fatty acid; Amino Acid Metabolism - valine, leucine and isoleucine degradation; Amino Acid Metabolism - tryptophan; EC 1.1.1.35 Chromosomal Location of Human Ortholog: 4q22-q26 Cellular Component: nucleoplasm; mitochondrion; mitochondrial matrix; mitochondrial inner membrane; cytoplasm Molecular Function: 3-hydroxyacyl-CoA dehydrogenase activity Biological Process: response to drug; fatty acid beta-oxidation; response to activity; cellular lipid metabolic process; negative regulation of insulin secretion; response to insulin stimulus Disease: 3-hydroxyacyl-coa Dehydrogenase Deficiency; Hyperinsulinemic Hypoglycemia, Familial, 4
Research Articles on HADH	**1.** Next-generation sequencing reveals deep intronic cryptic ABCC8 and HADH splicing founder mutations causing hyperinsulinism by pseudoexon activation.
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