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Rabbit Anti-HADHSC/Cy5 Conjugated antibody

SL3661R-Cy5

Product Name	Anti-HADHSC/Cy5
Chinese Name	Cy5 标记的短链 L-3 羟烷基辅酶 A 脱氢酶抗体
Alias	HAD; HADH; HADH1; HADHSC; HCDH; HCDH_MOUSE; HCDH_HUMAN; HHF4; Hydroxyacyl CoA dehydrogenase; Hydroxyacyl-coenzyme A dehydrogenase; hydroxyacyl-coenzyme A dehydrogenase, mitochondrial; L 3 hydroxyacyl Coenzyme A dehydrogenase short chain; M SCHAD; Medium and short chain L 3 hydroxyacyl coenzyme A dehydrogenase; Medium and short-chain L-3-hydroxyacyl-coenzyme A dehydrogenase; MGC8392; mitochondrial; MSCHAD; OTTHUMP00000162626; OTTHUMP00000219688; SCHAD; Short chain 3 hydroxyacyl CoA dehydrogenase mitochondrial; short chain 3-hydroxyacyl-coa dehydrogenase; Short-chain 3-hydroxyacyl-CoA dehydrogenase.
Research Area	immunology transcriptional regulatory factor Kinases and Phosphatases Diabetes Mitochondrion
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	(predicted:Mouse,Rat)
Applications	IF=1:100-500not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	35kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from mouse HADHSC
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The

lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Product Detail

background:
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-chain3-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.]

Function:
Plays an essential role in the mitochondrial beta-oxidation of short chain fatty acids. Exerts it highest activity toward 3-hydroxybutyryl-CoA.

Subunit:
Homodimer.

Subcellular Location:
Mitochondrion matrix.

Tissue Specificity:
Expressed in liver, kidney, pancreas, heart and skeletal muscle.

DISEASE:
Defects in HADH are the cause of 3-alpha-hydroxyacyl-CoA dehydrogenase deficiency (HADH deficiency) [MIM:231530]. 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) [MIM:609975]; 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.

Similarity:
Belongs to the 3-hydroxyacyl-CoA dehydrogenase family.

Database links:

Entrez Gene: 3033 Human

Entrez Gene: 15107 Mouse

Entrez Gene: 113965 Rat

Omim: 601609 Human

SwissProt: Q16836 Human

SwissProt: Q61425 Mouse

SwissProt: Q9WVK7 Rat

Unigene: 438289 Human

Unigene: 260164 Mouse

Unigene: 92789Rat

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

HADHSC 的缺少可导致家族性胰岛素过多低血糖综合症。