短链 L-3 羟烷基辅酶 A 脱氢酶抗体

产品货号： mIR3661

英文名称： HADHSC

中文名称： 短链 L-3 羟烷基辅酶 A 脱氢酶抗体

别　　名： 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.

研究领域： 免疫学　转录调节因子　激酶和磷酸酶　糖尿病　线粒体

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应　： Mouse, Rat,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 35kDa

细胞定位： 细胞浆 线粒体

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from mouse HADHSC:251-314/314

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍： 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.

SWISS:

Q61425

Gene ID:

15107

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。