abcam

Product datasheet

Anti-HADH antibody [1A12BC8] ab110284

4 图像

概述

产品名称	Anti-HADH抗体[1A12BC8]
描述	小鼠单克隆抗体[1A12BC8] to HADH
宿主	Mouse
经测试应用	**适用于:** IHC-P, ICC/IF, IP, Flow Cyt
种属反应性	**与反应:** Human
免疫原	Tissue, cells or virus. This information is considered to be commercially sensitive.
阳性对照	MRC5 fibroblasts; Human cerebellum tissue; Human liver mitochondria; HL-60 cells.
常规说明	This antibody clone is manufactured by Abcam. If you require a custom buffer formulation or conjugation for your experiments, please contact **orders@abcam.com**. The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As Product was previously marketed under the MitoSciences sub-brand.

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C. Do Not Freeze.
存储溶液	pH: 7.5 Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline
纯度	Proprietary Purification
纯化说明	Purity near homogeneity as judged by SDS-PAGE (purity >95%). The antibody was produced in vitro using hybridomas grown in serum-free medium, and then purified by biochemical fractionation.
克隆	单克隆

克隆编号	1A12BC8
同种型	IgG1
轻链类型	kappa

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab110284于以下的经测试应用**

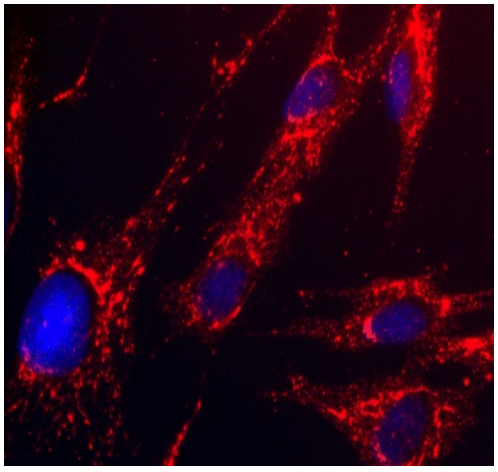
"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
IHC-P		1/250. Perform heat mediated antigen retrieval - 1 min pressure cook in 1mmol EDTA pH8.
ICC/IF		Use a concentration of 5 µg/ml.
IP		Use at an assay dependent concentration.
Flow Cyt		Use a concentration of 1 µg/ml. **ab170190** - Mouse monoclonal IgG1, is suitable for use as an isotype control with this antibody.

靶标

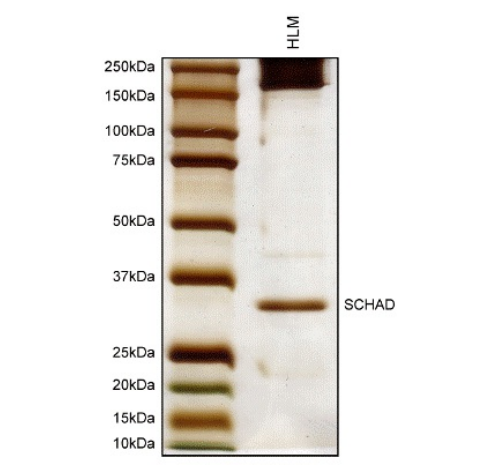
功能	Plays an essential role in the mitochondrial beta-oxidation of short chain fatty acids. Exerts it highest activity toward 3-hydroxybutyryl-CoA.
组织特异性	Expressed in liver, kidney, pancreas, heart and skeletal muscle.
通路	Lipid metabolism; fatty acid beta-oxidation.
疾病相关	Defects in HADH are the cause 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.
序列相似性	Belongs to the 3-hydroxyacyl-CoA dehydrogenase family.
细胞定位	Mitochondrion matrix.

图片



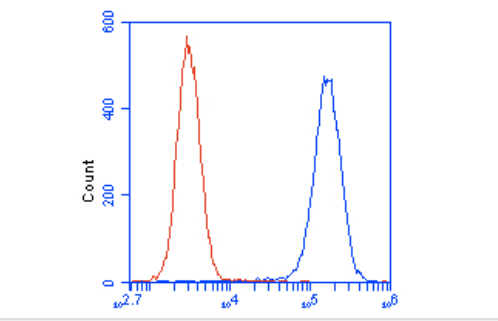
MRC5 fibroblasts labeled with ab110284 at 5µg/ml and stained with Texas Red conjugated to goat anti-mouse secondary.

Immunocytochemistry/ Immunofluorescence - Anti-HADH antibody [1A12BC8] (ab110284)



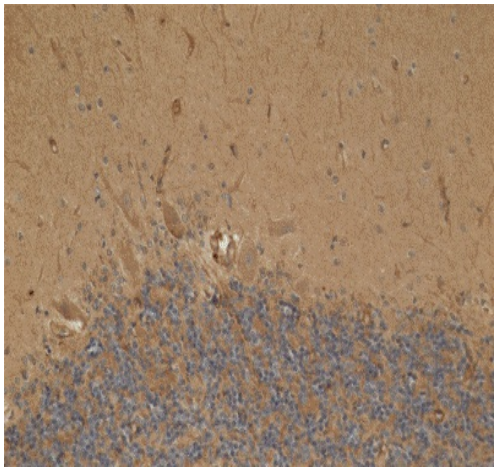
HADH immunocaptured from Human liver mitochondria using ab110284.

Immunoprecipitation - Anti-HADH antibody [1A12BC8] (ab110284)



HL-60 cells were stained with 1 µg/ml ab110284 (blue) or an equal amount of an isotype control antibody (red) and analyzed by flow cytometry.

Flow Cytometry - Anti-HADH antibody [1A12BC8] (ab110284)



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-HADH antibody [1A12BC8] (ab110284)

ab110284, at 1/250 dilution, staining HADH in formalin-fixed, paraffin-embedded Human cerebellum by Immunohistochemistry. HADH immunoactivity is most intense in neuronal cell bodies, most notably in the large Purkinje cells.

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