abcam

Product datasheet

Anti-HADH antibody [1A12BC8] ab110284

画像数 4

製品の概要

製品名 Anti-HADH antibody [1A12BC8]

製品の詳細 Mouse monoclonal [1A12BC8] to HADH

由来種 Mouse

アプリケーション 適用あり: IHC-P, ICC/IF, IP, Flow Cyt

種交差性 交差種: Human

免疫原 Tissue, cells or virus. This information is considered to be commercially sensitive.

ポジティブ・コントロール MRC5 fibroblasts; Human cerebellum tissue; Human liver mitochondria; HL-60 cells.

特記事項

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.

製品の特性

製品の状態 Liquid

保存方法 Shipped at 4°C. Store at +4°C. Do Not Freeze.

バッファー pH: 7.5

Preservative: 0.02% Sodium azide Constituent: HEPES buffered saline

精製度 Proprietary Purification

特記事項(精製) 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.

ポリ/モノ モノクローナル

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クローン名 1A12BC8

アイソタイプ lgG1 軽鎖の種類 kappa

アプリケーション

The Abpromise guarantee <u>Abpromise保証は、</u>次のテスト済みアプリケーションにおけるab110284の使用に適用されます アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
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. <u>ab170190</u> - Mouse monoclonal lgG1, is suitable for use as an isotype control with this antibody.

ターゲット情報

機能 Plays an essential role in the mitochondrial beta-oxidation of short chain fatty acids. Exerts it

highest activity toward 3-hydroxybutyryl-CoA.

組織特異性 Expressed in liver, kidney, pancreas, heart and skeletal muscle.

パスウェイ Lipid metabolism; fatty acid beta-oxidation.

関連疾患 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

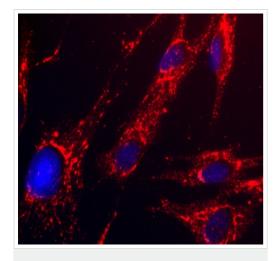
and is due to defective negative feedback regulation of insulin secretion by low glucose levels. It causes nesidioblastosis, a diffuse abnormality of the pancreas in which there is extensive, often disorganized formation of new islets. Unless early and aggressive intervention is undertaken, brain damage 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.

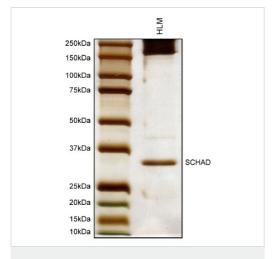
配列類似性 Belongs to the 3-hydroxyacyl-CoA dehydrogenase family.

細胞内局在 Mitochondrion matrix.

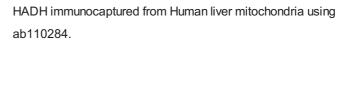


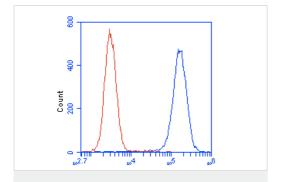
Immunocytochemistry/ Immunofluorescence - Anti-HADH antibody [1A12BC8] (ab110284)

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



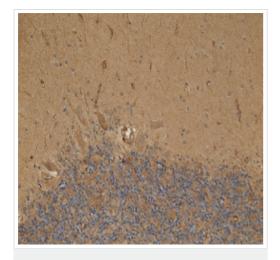
Immunoprecipitation - Anti-HADH antibody [1A12BC8] (ab110284)





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

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



Immunohistochemistry (Formalin/PFA-fixed paraffinembedded 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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