abcam

Product datasheet

Anti-HADHA antibody ab54477

3 Abreviews 44 References 1 图像

概述

产品名称	Anti-HADHA抗体
描述	**兔多克隆抗体**to HADHA
宿主	Rabbit
经测试应用	**适用于:** WB
种属反应性	**与反应:** Mouse, Rat, Human
免疫原	Synthetic peptide surrounding amino acid 750 (Human)
阳性对照	Jurkat cell lysate 3T3 cell lysate Rat kidney lysate
常规说明	This product is manufactured by BioVision, an Abcam company and was previously called 3721 TFP1/HADHA Antibody. 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

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: 0.01% Thimerosal (merthiolate) Constituents: 0.5% BSA, 30% Glycerol (glycerin, glycerine), PBS
纯度	Protein A purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab54477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
WB	★★★★★ (3)	Use a concentration of 0.5 - 4 µg/ml. Detects a band of approximately 83 kDa (predicted molecular weight: 83 kDa).

靶标

功能 Bifunctional subunit.

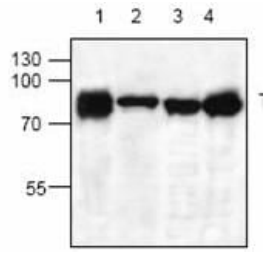
通路 Lipid metabolism; fatty acid beta-oxidation.

疾病相关 Defects in HADHA are a cause of trifunctional protein deficiency (TFP deficiency) [MIM:609015]. The clinical manifestations are very variable and include hypoglycemia, cardiomyopathy and sudden death. Phenotypes with mainly hepatic and neuromyopathic involvement can also be distinguished. Biochemically, TFP deficiency is defined by the loss of all enzyme activities of the TFP complex.
Defects in HADHA are the cause of long-chain 3-hydroxyl-CoA dehydrogenase deficiency (LCHAD deficiency) [MIM:609016]. The clinical features are very similar to TFP deficiency. Biochemically, LCHAD deficiency is characterized by reduced long-chain 3-hydroxyl-CoA dehydrogenase activity, while the other enzyme activities of the TFP complex are normal or only slightly reduced.
Defects in HADHA are a cause of maternal acute fatty liver of pregnancy (AFLP) [MIM:609016]. AFLP is a severe maternal illness occurring during pregnancies with affected fetuses. This disease is associated with LCHAD deficiency and characterized by sudden unexplained infant death or hypoglycemia and abnormal liver enzymes (Reye-like syndrome).

序列相似性 In the N-terminal section; belongs to the enoyl-CoA hydratase/isomerase family.
In the central section; belongs to the 3-hydroxyacyl-CoA dehydrogenase family.

细胞定位 Mitochondrion.

图片



Western blot - Anti-HADHA antibody (ab54477)

All lanes : Anti-HADHA antibody (ab54477) at 4 µg/ml

Lanes 1-2 : Jurkat cell lysate 30-50 ug/lane.
Lane 3 : 3T3 cell lysate 30-50 ug/lane.
Lane 4 : Rat kidney lysate 30-50 ug/lane.

Secondary
All lanes : Anti-Rabbit IgG, HRP-Linked Antibody at 1/5000 dilution

Predicted band size: 83 kDa
Observed band size: 83 kDa

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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- Replacement or refund for products not performing as stated on the datasheet
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