abcam

Product datasheet

Anti-HADHA antibody ab54477

3 Abreviews 44 References 1 Image

Overview

Product name	Anti-HADHA antibody
Description	Rabbit polyclonal to HADHA
Host species	Rabbit
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Mouse, Rat, Human
Immunogen	Synthetic peptide surrounding amino acid 750 (Human)
Positive control	Jurkat cell lysate 3T3 cell lysate Rat kidney lysate
General notes	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

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	Preservative: 0.01% Thimerosal (merthiolate) Constituents: 0.5% BSA, 30% Glycerol (glycerin, glycerine), PBS
Purity	Protein A purified
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab54477 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB	★★★★★ (3)	Use a concentration of 0.5 - 4 µg/ml. Detects a band of approximately 83 kDa (predicted molecular weight: 83 kDa).

Target

Function Bifunctional subunit.

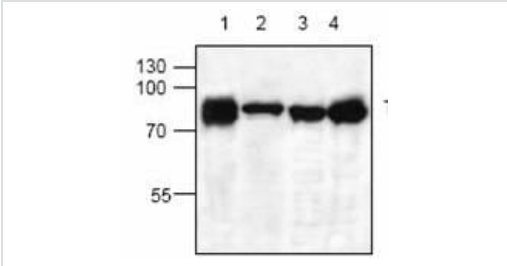
Pathway Lipid metabolism; fatty acid beta-oxidation.

Involvement in disease 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).

Sequence similarities 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.

Cellular localization Mitochondrion.

Images



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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