

Product datasheet

Recombinant Human HADHA protein ab158631

[1 Image](#)

Description

Product name	Recombinant Human HADHA protein
Expression system	Wheat germ
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human

Sequence

MVACRAIGILSRFSAFRILRSRGYICRNFTGSSALLTRTHINY
GVKGDVA
VVRINSPNSKVNLSKELHSEFSEVMNEIWASDQIRSAVLI
SSKPGCFIA
GADINMLAACKTLQEVTQLSQEAQRVEKLEKSTKPIVAAIN
GSCLGGGL
EVAISCQYRIATKDRKTVLGTPEVLLGALPGAGGTQRLPK
MVGVPAALDM
MLTGRSIRADRAKKMGLVDQLVEPLGPGPKPPEERTIEYL
EEVAITFAKG
LADKKISPKRDKGLVEKLTAYAMTIPFVRQQVYKKVEEKV
RKQTKGLYPA
PLKIIDVVKTGIEQGSDAGYLCESQKFGELVMTKESKALM
GLYHGQVLCK
KNKFGAPQKDVKHLAILGAGLMGAGIAQVSVDKGLKTILK
DATLTALDRG
QQQVFKGLNDKVKKKALTSFERDSIFSNTLGQLDYQGFEK
ADMVIEAVFE
DLSLKHRVLKEVEAVIPDHCIFASNTSALPISEIAAVSKRPE
KVIGMHYF
SPVDKMQLLEIITTEKTSKDTASAVAVGLKQKVIIVKD
GPGFYTTRC
LAPMMSEVIRILQEGVDPKKLDSLTSFGFPVGAATLVDE
VGVDVAKHVA
EDLGKVFGERFGGGNPELLTQMVSKGFLGRKSGKGFYIY
QEGVKRKDLNS
DMSILASLKLPPKSEVSSDEDIQFRLVTRFVNEAVMCLQ
EGILATPAEG
DIGAVFGLGFPCLGGPFRFVDLYGAQKIMDRLKKYEAAY

Amino acids	1 to 763
Tags	GST tag N-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab158631** in the following tested applications.

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

Applications	Western blot
	ELISA
Form	Liquid
Additional notes	

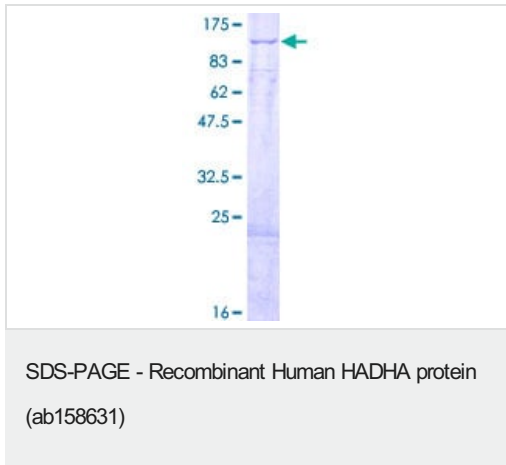
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl
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General Info

Function	Bifunctional subunit.
Pathway	Lipid metabolism; fatty acid beta-oxidation.
Involvement in disease	<p>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.</p> <p>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.</p> <p>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).</p>
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



ab158631 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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

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