

Recombinant Human HADHA protein ab158631

1 图像

描述	
产品名称	重组人HADHA蛋白
表达系统	Wheat germ
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MVACRAIGILSRFSAFRILRSRGYICRNFTGSSALLTRTHIN YGVKGDVA VVRINSPNSKVNTLSKELHSEFSEVMNEIWASDQIRSAVLIS SKPGCFIA GADINMLAACKTLQEVTQLSQEAQRIVEKLEKSTKPIVAAIN GSCLGGGL EVAISCQYRIATKDRKTVLGTPEVLLGALPGAGGTQRLPKMV GVPAALDM MLTGRSIRADRAKKMGLVDQLVEPLGPGLKPPEERTIEYLEE VAITFAKG LADKKISPKRDKGLVEKLTAYAMTIPFVRQQVYKKVEEKVRK QTKGLYPA PLKIIDVVKTGIEQGSDAGYLCESQKFGELVMTKESKALMGL YHGQVLCK KNKFGAPQKDVKHLAILGAGLMGAGIAQVSVDKGLKTILKDA TLTALDRG QQQVFKGLNDKVKKKALTSFERDSIFSNLTGQLDYQGFEKAD MVEAVFE DLSLKHRVLKEVEAIPDHCIFASNTSALPISEIAAVSKRPE KVIGMHYF SPVDKMQLLEIITTEKTSKDTSSASAVAVGLKQGKVIIIVKDG PGFYTTTC LAPMMSEVIRILQEGVDPKKLDSLTTSGFPVGAATLVDEVG VDVAKHVA EDLGKVFGERFGGNGPELLTQMVSKGFLGRKSGKGFYIYQEG VKRKDLNS DMDSILASLKLPPKSEVSSDEDIQFRLVTRFVNEAVMCLQEG ILATPAEG DIGAVFGLGFPPCLGGPFRFVDLYGAQKIVDRLKKYEAAYGK

氨基酸	1 to 763
标签	GST tag N-Terminus

技术指标

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.

应用	Western blot
	ELISA
形式	Liquid
补充说明	

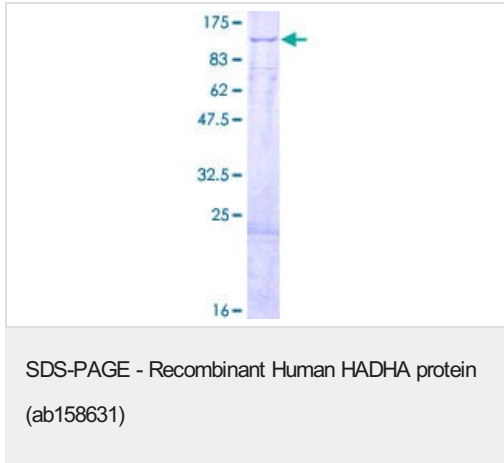
制备和贮存

稳定性和存储	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
--------	--

常规信息

功能	Bifunctional subunit.
通路	Lipid metabolism; fatty acid beta-oxidation.
疾病相关	<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>
序列相似性	<p>In the N-terminal section; belongs to the enoyl-CoA hydratase/isomerase family.</p> <p>In the central section; belongs to the 3-hydroxyacyl-CoA dehydrogenase family.</p>
细胞定位	Mitochondrion.

图片



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"

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.cn/abpromise> or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors