abcam

Product datasheet

Recombinant Human CPT2 protein ab114539

1 图像

描述

产品名称	重组人CPT2蛋白
表达系统	Wheat germ
Accession	P23786
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MVPRLLLRAW PRGPAVGPGA PSRPLSAGSG PGQYLQRSIV PTMHYQDSLP RLPIPKLEDT IRRYLSAQKP LLNDGQFRKT EQFCKSFENG IGKELHEQLV ALDKQNKHTS YISGPWFDMY LSARDSVVLN FNPFMAFNPD PKSEYNDQLT RATNMTVSAI RFLKTLRAGL LEPEVFHLNP AKSDTITFKR LIRFVPSSLS WYGAYLVNAY PLDMSQYFRL FNSTRLPKPS RDELFTDDKA RHLLVLRKGN FYIFDVLDQD GNIVSPSEIQ AHLKYILSDS SPAPEFPLAY LTSENRDIWA ELRQKLMSSG NEESLRKVDS AVFCLCLDDF PIKDLVHLSH NMLHGDGTNR WFDKSFNLII AKDGSTAVHF EHSWGDGVAV LRFFNEVFKD STQTPAVTPQ SQPATTDSTV TVQKLNFELT DALKTGITAA KEKFDATMKT LTIDCVQFQR GGKEFLKKQK LSPDAVAQLA FQMAFLRQYG QTVATYESCS TAAFKHGRTE TIRPASVYTK RCSEAFVREP SRHSAGELQQ MMVECSKYHG QLTKEAAMGQ GFDRHLFALR HLAAAKGIIL PELYLDPAYG QINHNVLSTS TLSSPAVNLG GFAPVVSDGF GVGYAVHDNW IGCNVSSYPG RNAREFLQCV EKALEDMFDA LEGKSIKS
预测分子量	101 kDa including tags
氨基酸	1 to 658

技术指标

Our **Abpromise guarantee** covers the use of **ab114539** in the following tested applications.

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

应用	SDS-PAGE ELISA Western blot
形式	Liquid

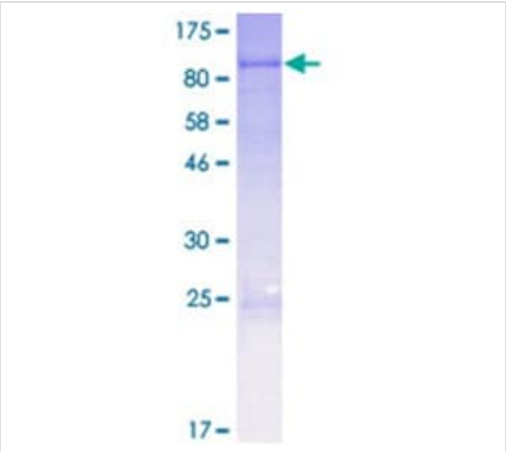
制备和贮存

稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

常规信息

通路	Lipid metabolism; fatty acid beta-oxidation.
疾病相关	Defects in CPT2 are the cause of carnitine palmitoyltransferase 2 deficiency (CPT2D) [MIM:255110, 600649]; also known as CPT-II deficiency or CPT2 deficiency. CPT2D is an autosomal recessive disorder characterized by recurrent myoglobinuria, episodes of muscle pain, stiffness, and rhabdomyolysis. These symptoms are triggered by prolonged exercise, fasting or viral infection and patients are usually young adults. In addition to this classical, late-onset, muscular type, a hepatic or hepatocardiomuscular form has been reported in infants. Clinical pictures in these children or neonates include hypoketotic hypoglycemia, liver dysfunction, cardiomyopathy and sudden death. Defects in CPT2 are the cause of carnitine palmitoyltransferase 2 deficiency, lethal neonatal (CPT2D-LN) [MIM:608836]; also known as lethal neonatal CPT-II deficiency. It is a lethal neonatal form of CPT2D. This rarely presentation is antenatal with cerebral periventricular cysts and cystic dysplastic kidneys. The clinical variability of the disease is likely attributed to the variable residual enzymatic activity.
序列相似性	Belongs to the carnitine/choline acetyltransferase family.
细胞定位	Mitochondrion inner membrane.

图片



SDS-PAGE - Recombinant Human CPT2 protein (ab114539)

ab114539 anaylsed 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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