

Recombinant Human CPT2 protein ab114539

1 Image

Description

Product name	Recombinant Human CPT2 protein
Expression system	Wheat germ
Accession	P23786
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MVPRLLLRRAW PRGPAVGPGA PSRPLSAGSG PGQYLQRSIV PTMHYQDQLP RLPIPKLEDT IRRYLSAQKP LLNDGQFRKT EQFCKSFENG IGKELHEQLV ALDKQNKHTS YISGPWFDMY LSARDSVVLN FNPFMAFNPD PKSEYNDQLT RATNMTVSAI RFLKTLRAGL LEPEVFHLNP AKSDTITFKR LIRFVPSSLS WYGAYLVNAY PLDMSQYFRL FNSTRLPKPS RDELFTDDKA RHLLVLRKGN FYIFDVLDQD GNVSPSEIQ AHLKYILSDS SPAPEFPLAY LTSENARDIWA ELRQKLMSSG NEESLRKVDS AVFCLCLDDF PIKDLVHLSH NMLHGDGTNR WFDKSFNLII AKDGSTAVHF EHSWGDGVAV LRFFNEVFKD STQTPAVTPQ SQPATTDSTV TVQKLNFEEL DALKTGITAA KEKFDATMKT LTIDCVQFQR GGKEFLKKQK LSPDAVAQLA FQMAFLRQYG QTVATYESCS TAAFKHGRTE TIRPASVYTK RCSEAFVREP SRHSAGELQQ MMVECSKYHG QLTKEAAMGQ GFDRHLFALR HLAAAKGIIL PELYLDPAYG QINHNVLSTS TLSSPAVNLG GFAPVVSDGF GVGAVVHDNW IGCNVSSYPG RNAREFLQCV EKALEDMFDA LEGKSIKS
Predicted molecular weight	101 kDa including tags
Amino acids	1 to 658

Specifications

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.

Applications	SDS-PAGE
	ELISA
	Western blot
Form	Liquid

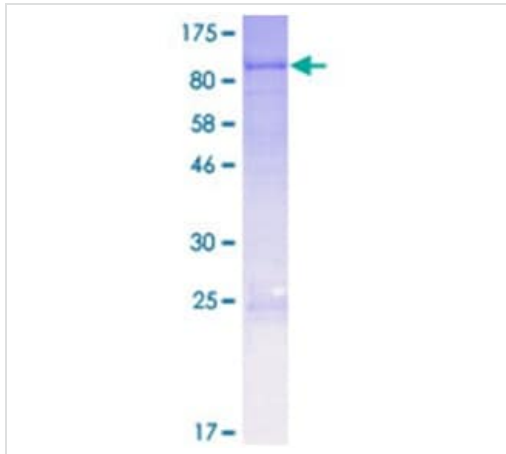
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl
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General Info

Pathway	Lipid metabolism; fatty acid beta-oxidation.
Involvement in disease	<p>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 hepatocardiomyopathy form has been reported in infants. Clinical pictures in these children or neonates include hypoketotic hypoglycemia, liver dysfunction, cardiomyopathy and sudden death.</p> <p>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.</p>
Sequence similarities	Belongs to the carnitine/choline acetyltransferase family.
Cellular localization	Mitochondrion inner membrane.

Images



SDS-PAGE - Recombinant Human CPT2 protein
(ab114539)

ab114539 analysed on a 12.5% SDS-PAGE Stained with Coomassie Blue.

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