# abcam

## Product datasheet

# Recombinant Human CPT2 protein ab114539

### 画像数1

#### 製品の詳細

製品名 Recombinant Human CPT2 protein

発現系Wheat germアクセッション番号P23786

タンパク質長 Full length protein

Animal free No

**由来** Recombinant

生物種 Human

配列 MVPRLLLRAW PRGPAVGPGA PSRPLSAGSG

PGQYLQRSIV PTMHYQDSLP RLPIPKLEDT IRRYLSAQKP LLNDGQFRKT EQFCKSFENG

IGKELHEQLV ALDKQNKHTS YISGPWFDMY

TORCETTE ALDRONAMITS TISOFWI DATE

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

SIQITAVITQ SQLATTUSTV TVQKEM EET

DALKTGITAA KEKFDATMKT LTIDCVQFQR

GGKEFLKKQK LSPDAVAQLA FQMAFLRQYG

QTVATYESCS TAAFKHGRTE TIRPASVYTK

RCSEAFVREP SRHSAGELQQ MMVECSKYHG QLTKEAAMGQ GFDRHLFALR HLAAAKGIIL

PELYLDPAYG QINHNVLSTS TLSSPAVNLG

GFAPVVSDGF GVGYAVHDNW IGCNVSSYPG

RNAREFLQCV EKALEDMFDA LEGKSIKS

予測される分子量 101 kDa including tags

**領域** 1 to 658

特性

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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.

アプリケーション SDS-PAGE

**ELISA** 

Western blot

製品の状態 Liquid

#### 前処理および保存

保存方法および安定性 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 HCI

#### 関連情報

パスウェイ Lipid metabolism; fatty acid beta-oxidation.

**関連疾患**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.

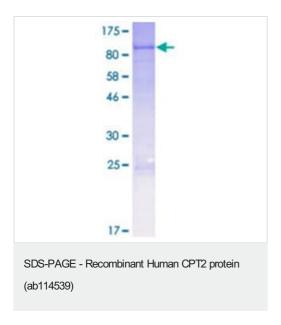
Defects in CPT2 are the cause of carnitine palmitoyltransferase 2 deficiency, lethal neonatal (CPT2D-LN) [MIM:608836]; also known as lethal neonatal CPT-II deficiency. It is a lethal neonatal form of CPT2D. This rarely presentation is antenatal with cerebral periventricular cysts and cystic dysplastic kidneys. The clinical variability of the disease is likely attributed to the variable residual

enzymatic activity.

**配列類似性** Belongs to the carnitine/choline acetyltransferase family.

細胞内局在 Mitochondrion inner membrane.

#### 画像



ab114539 analysed 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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