

Anti-CPT2 antibody ab54589

2 图像

概述

产品名称	Anti-CPT2抗体
描述	小鼠单克隆抗体to CPT2
宿主	Mouse
经测试应用	适用于: WB
种属反应性	与反应: Recombinant fragment 预测可用于: Human 
免疫原	Recombinant fragment: WFDKSFNLI AKDGSTAIHF EHSWGDGVAV LRFNEVFKD STQTPAVTPQ SQPATTSTV TVQKLNFEELT DALKTGITAA KEKFDATMKT LTIDCVQFQR , corresponding to amino acids 351-451 of Human CPT2 Run BLAST with ExPASy Run BLAST with NCBI

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: None PBS, pH 7.2
纯度	Protein G purified
克隆	单克隆
同种型	IgG2b
轻链类型	kappa

应用

The Abpromise guarantee **Abpromise™** 承诺保证使用ab54589于以下的经测试应用

“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明

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WB		Use a concentration of 1 - 5 µg/ml. This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.

靶标

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 hepatocardiomyopathy 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.

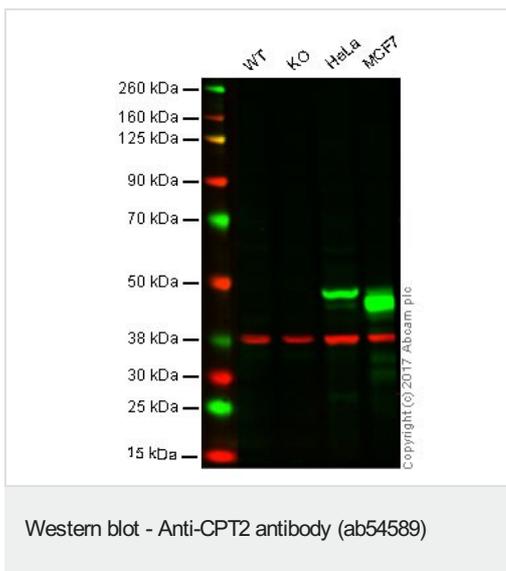
序列相似性

Belongs to the carnitine/choline acetyltransferase family.

细胞定位

Mitochondrion inner membrane.

图片



Lane 1: Wild type HAP1 whole cell lysate (20 µg)

Lane 2: CPT2 knockout HAP1 whole cell lysate (20 µg)

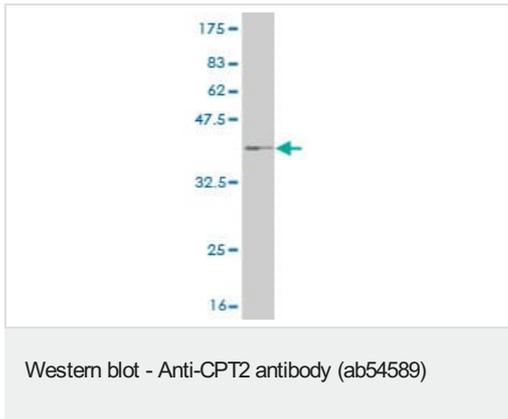
Lane 3: HeLa whole cell lysate (20 µg)

Lane 4: MCF7 whole cell lysate (20 µg)

Lanes 1 - 4: Merged signal (red and green). Green - ab54589 observed at 70 kDa. Red - loading control, **ab181602**, observed at 37 kDa.

ab54589 was shown not to specifically react with CPT2 when CPT2 knockout samples were used. Wild-type and CPT2 knockout samples were subjected to SDS-PAGE. Ab54589 and **ab181602** (Rabbit anti GAPDH loading control) were incubated overnight at 4°C at 1 µg/ml and 1/10000 dilution respectively. Blots were developed with Goat anti-Mouse IgG H&L (IRDye® 800CW) preabsorbed **ab216772** and Goat anti-Rabbit IgG H&L (IRDye® 680RD) preabsorbed **ab216777** secondary antibodies at 1/20000

dilution for 1 hour at room temperature before imaging.



Western blot against tagged recombinant protein immunogen using ab54589 CPT2 antibody at 1ug/ml. Predicted band size of immunogen is 37 kDa.

This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.

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