abcam

Product datasheet

Recombinant Human MCCC2 protein ab163770

1 Image

Overview

Product name	Recombinant Human MCCC2 protein
Protein length	Full length protein

Description

Nature	Recombinant
Source	Wheat germ
Amino Acid Sequence	
Species	Human
Sequence	MWAVLRLALRPCARASPAGPRAYHGDSVASLGTQPD LGSALYQENYKQMK ALVNQLHERVEHIKLGGGEKARALHISRGKLLPRERIDN LIDPGSPFLEL SQFAGYQLYDNEEVPGGGIITGIGRVSGVECMIIANDAT VKGGAYYPVTV KKQLRAQEIAMQNRLPCIYLVDSGGAYLPRQADVFPD RDHFGRTFYNQAI MSSKNIAQIAVVMGSCTAGGAYVPAMADENIIVRKQGTI FLAGPPLVKAA TGEEVSAEDLGGADLHCRKSGVSDHWALDDHHALHL TRKVVRNLNYQKKL DVTIEPSEEPLFPADELYGIVGANLKRSFDVREVIARIV DGSRFTEFKAF YGDTLVTGFARIFGYPVGIVGNNGVLFSESAKKGTHFV QLCCQRNIPLLF LQNITGFMVGREYEAEGIAKDGAKMVAAVACAQVPKIT LIIGGSYGAGNY GMCGRAYSPRFLYIWPNARISVMGGEQAANVLATITKD QRAREGKQFSSA DEAALKEPIIKKFEEEGNPYYSSARVWDDGIIDPADTRL VLGLSFSAALN APIEKTDFGIFRM
Amino acids	1 to 563
Tags	GST tag N-Terminus

Specifications

Our Abpromise guarantee covers the use of **ab163770** in the following tested applications.

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

Applications	ELISA Western blot
Form	Liquid
Additional notes	Protein concentration is above or equal to 0.05 mg/ml.

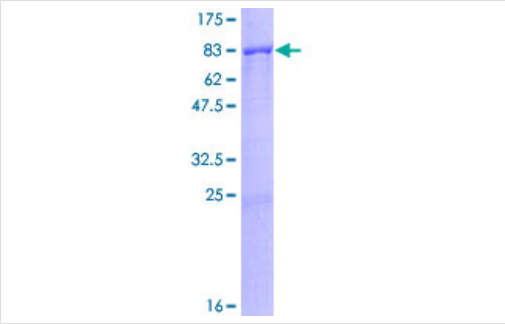
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

General Info

Pathway	Amino-acid degradation; L-leucine degradation; (S)-3-hydroxy-3-methylglutaryl-CoA from 3-isovaleryl-CoA: step 2/3.
Involvement in disease	Defects in MCCC2 are the cause of methylcrotonoyl-CoA carboxylase deficiency type 2 (MCC2 deficiency) [MIM:210210]. MCC2 deficiency is an autosomal recessive disorder of leucine catabolism. The phenotype is variable, ranging from neonatal onset with severe neurological involvement to asymptomatic adults. There is a characteristic organic aciduria with massive excretion of 3-hydroxyisovaleric acid and 3-methylcrotonylglycine, usually in combination with a severe secondary carnitine deficiency.
Sequence similarities	Belongs to the AccD/PCCB family. Contains 1 carboxyltransferase domain.
Cellular localization	Mitochondrion matrix.

Images



SDS-PAGE - Recombinant Human MCCC2 protein (ab163770)

ab163770 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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