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MCCC1 Antibody (monoclonal) (M01)

Mouse monoclonal antibody raised against a partial recombinant MCCC1.
Catalog # AT2816a

Product Information

Application	WB, E
Primary Accession	Q96RQ3
Other Accession	NM_020166
Reactivity	Human
Host	mouse
Clonality	monoclonal
Isotype	IgG2a Kappa
Clone Names	2G8
Calculated MW	80473

Additional Information

Gene ID	56922
Other Names	Methylcrotonoyl-CoA carboxylase subunit alpha, mitochondrial, MCCase subunit alpha, 3-methylcrotonyl-CoA carboxylase 1, 3-methylcrotonyl-CoA carboxylase biotin-containing subunit, 3-methylcrotonyl-CoA:carbon dioxide ligase subunit alpha, MCCC1, MCCA
Target/Specificity	MCCC1 (NP_064551, 526 a.a. ~ 625 a.a) partial recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Dilution	WB~~1:500~1000 E~~N/A
Format	Clear, colorless solution in phosphate buffered saline, pH 7.2 .
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.
Precautions	MCCC1 Antibody (monoclonal) (M01) is for research use only and not for use in diagnostic or therapeutic procedures.

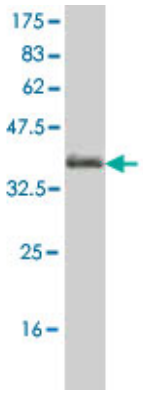
Background

This gene encodes the large subunit of 3-methylcrotonyl-CoA carboxylase. This enzyme functions as a heterodimer and catalyzes the carboxylation of 3-methylcrotonyl-CoA to form 3-methylglutaconyl-CoA. Mutations in this gene are associated with 3-Methylcrotonylglycinuria, an autosomal recessive disorder of leucine catabolism.

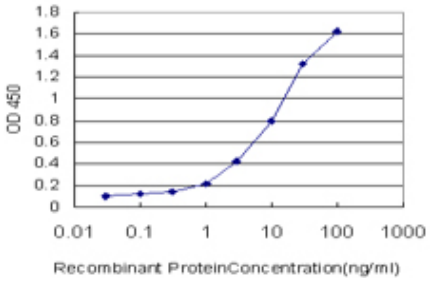
References

Personalized smoking cessation: interactions between nicotine dose, dependence and quit-success genotype score. Rose JE, et al. Mol Med, 2010 Jul-Aug. PMID 20379614.Cryptic exon activation by disruption of exon splice enhancer: novel mechanism causing 3-methylcrotonyl-CoA carboxylase deficiency. Stucki M, et al. J Biol Chem, 2009 Oct 16. PMID 19706617.3-Methylcrotonyl-CoA carboxylase deficiency: phenotypic variability in a family. Eminoglu FT, et al. J Child Neurol, 2009 Apr. PMID 19339287.Toward a confocal subcellular atlas of the human proteome. Barbe L, et al. Mol Cell Proteomics, 2008 Mar. PMID 18029348.Novel mutations in five Japanese patients with 3-methylcrotonyl-CoA carboxylase deficiency. Uematsu M, et al. J Hum Genet, 2007. PMID 17968484.

Images



Antibody Reactive Against Recombinant Protein.Western Blot detection against Immunogen (36.74 KDa) .



Detection limit for recombinant GST tagged MCCC1 is approximately 0.1ng/ml as a capture antibody.