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HMGCL Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP18139a

Product Information

Application	WB, E
Primary Accession	P35914
Other Accession	NP_000182.2
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB22965
Calculated MW	34360
Antigen Region	71-99

Additional Information

Gene ID	3155
Other Names	Hydroxymethylglutaryl-CoA lyase, mitochondrial, HL, HMG-CoA lyase, 3-hydroxy-3-methylglutarate-CoA lyase, HMGCL
Target/Specificity	This HMGCL antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 71-99 amino acids from the N-terminal region of human HMGCL.
Dilution	WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	HMGCL Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	HMGCL
Function	Mitochondrial 3-hydroxy-3-methylglutaryl-CoA lyase that catalyzes a cation-dependent cleavage of (S)-3-hydroxy-3- methylglutaryl-CoA into acetyl-CoA and acetoacetate, a key step in ketogenesis. Terminal step in

	leucine catabolism. Ketone bodies (beta- hydroxybutyrate, acetoacetate and acetone) are essential as an alternative source of energy to glucose, as lipid precursors and as regulators of metabolism.
Cellular Location	Mitochondrion matrix {ECO:0000250|UniProtKB:P38060}. Peroxisome {ECO:0000250|UniProtKB:P38060}. Note=Unprocessed form is peroxisomal {ECO:0000250|UniProtKB:P38060}
Tissue Location	Highest expression in liver. Expressed in pancreas, kidney, intestine, testis, fibroblasts and lymphoblasts. Very low expression in brain and skeletal muscle. The relative expression of isoform 2 (at mRNA level) is highest in heart (30%), skeletal muscle (22%), and brain (14%).

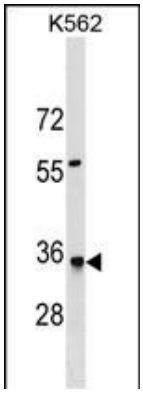
Background

The protein encoded by this gene belongs to the HMG-CoA lyase family. It is a mitochondrial enzyme that catalyzes the final step of leucine degradation and plays a key role in ketone body formation. Mutations in this gene are associated with HMG-CoA lyase deficiency. Alternatively spliced transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq].

References

Fu, Z., et al. J. Biol. Chem. 285(34):26341-26349(2010)
Pierron, S., et al. Arch Pediatr 17(1):10-13(2010)
Menao, S., et al. Hum. Mutat. 30 (3), E520-E529 (2009) :
Lin, W.D., et al. Clin. Chim. Acta 401 (1-2), 33-36 (2009) :
Carrasco, P., et al. Mol. Genet. Metab. 91(2):120-127(2007)

Images



HMGCL Antibody (N-term) (Cat. #AP18139a) western blot analysis in K562 cell line lysates (35ug/lane).This demonstrates the HMGCL antibody detected the HMGCL protein (arrow).