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HMGCL Rabbit mAb

Catalog # AP75555

Product Information

Application	WB, IHC-P
Primary Accession	P35914
Reactivity	Human
Host	Rabbit
Clonality	Monoclonal Antibody
Isotype	IgG
Conjugate	Unconjugated
Purification	Affinity Purified
Calculated MW	34360

Additional Information

Gene ID	3155
Other Names	HMGCL
Dilution	WB~~1/500-1/1000 IHC-P~~N/A
Format	Liquid in 50mM Tris-Glycine(pH 7.4), 0.15M NaCl, 40%Glycerol, 0.01% sodium azide and 0.05% BSA.
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.

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.

Images