HMGCL Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	HMGCL, Polyclonal Antibody
Full Product Name	HMGCL, ID (HMGCL, Hydroxymethylglutaryl-CoA lyase, mitochondrial, 3-hydroxy-3-methylglutarate-CoA lyase) (MaxLight 490)
Product Gene Name	anti-HMGCL antibody
Product Synonym Names	HMGCL; Hydroxymethylglutaryl-CoA lyase, mitochondrial; 3-hydroxy-3-methylglutarate-CoA lyase
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with MaxLight490.
Purity/Purification	Purified by Protein A Affinity Chromatography.
Preparation and Storage	Store product at 4 degree C in the dark. DO NOT FREEZE! Stable at 4 degree C for 12 months after receipt as an undiluted liquid. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. Caution: MaxLight490 conjugates are sensitive to light. For maximum recovery of product, centrifuge the original vial prior to removing the cap.
Molecular Weight	20,222 Da
OMIM	phenotype 613898
3D Structure	ModBase 3D Structure for P35914
Immunogen	HMGCL antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 160-190 amino acids from the Central region of human HMGCL.
Conjugate	MaxLight490
Note	Preservative Free
Product Categories/Family for anti-HMGCL antibody	Antibodies; Mitochondrial Proteins
Applications Tested/Suitable for anti-HMGCL antibody	Western Blot (WB), FLISA
Application Notes for anti-HMGCL antibody	WB: 1:100-500 FLISA: 1:1,000 Applications are based on unconjugated antibody.
Related Product Information for anti-HMGCL antibody	HMGCL 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].
Product References and Citations for anti-HMGCL antibody	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).

Other Notes	Small volumes of anti-HMGCL antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for HMGCL purchase	MBS6307303 is an antibody targeting HMGCL. Also known as HMGCL, ID (HMGCL, Hydroxymethylglutaryl-CoA lyase, mitochondrial, 3-hydroxy-3-methylglutarate-CoA lyase) (MaxLight 490). Reactive in Human. Host: Rabbit.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for HMGCL. It may not necessarily be applicable to this product.

NCBI GI #	62198232
NCBI GeneID	3155
NCBI Accession #	NP_000182.2
NCBI Related Accession #	NP_001159531.1
UniProt Primary Accession #	P35914
UniProt Secondary Accession #	Q6IBC0; Q96FP8; B4DUP4; B7UCC6; D3Y5K7
UniProt Related Accession #	P35914
GenBank Nucleotide #	NM_000191.2
NCBI Official Full Name	hydroxymethylglutaryl-CoA lyase, mitochondrial isoform 1
NCBI Official Synonym Full Names	3-hydroxymethyl-3-methylglutaryl-CoA lyase
NCBI Official Symbol	HMGCL
NCBI Official Synonym Symbols	HL
UniProt Protein Name	Hydroxymethylglutaryl-CoA lyase, mitochondrial
UniProt Synonym Protein Names	3-hydroxy-3-methylglutarate-CoA lyase
UniProt Gene Name	HMGCL
UniProt Synonym Gene Names	HL; HMG-CoA lyase
UniProt Entry Name	HMGCL_HUMAN
NCBI Protein Information	hydroxymethylglutaryl-CoA lyase, mitochondrial; 3-hydroxy-3-methylglutarate-CoA lyase; 3-hydroxy-3-methylglutaryl-CoA lyase; 3-hydroxymethyl-3-methylglutaryl-Coenzyme A lyase; HMG-CoA lyase; hydroxymethylglutaricaciduria; mitochondrial 3-hydroxy-3-methylg
NCBI Summary for HMGCL	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, Oct 2009]

UniProt Comments for HMGCL	HMGCL: Key enzyme in ketogenesis (ketone body formation). Terminal step in leucine catabolism. Defects in HMGCL are the cause of 3-hydroxy-3- methylglutaryl-CoA lyase deficiency (HMGCLD); also known as hydroxymethylglutaricaciduria or HL deficiency. An autosomal recessive disease affecting ketogenesis and L-leucine catabolism. The disease usually appears in the first year of life after a fasting period and its clinical acute symptoms include vomiting, seizures, metabolic acidosis, hypoketotic hypoglycemia and lethargy. These symptoms sometimes progress to coma, with fatal outcome in some cases. Belongs to the HMG-CoA lyase family. 2 isoforms of the human protein are produced by alternative splicing. Protein type: Carbohydrate Metabolism - butanoate; Mitochondrial; Amino Acid Metabolism - valine, leucine and isoleucine degradation; EC 4.1.3.4; Lyase; Lipid Metabolism - synthesis and degradation of ketone bodies Chromosomal Location of Human Ortholog: 1p36.1-p35 Cellular Component: mitochondrion; mitochondrial matrix; mitochondrial inner membrane; peroxisome Molecular Function: hydroxymethylglutaryl-CoA lyase activity; protein homodimerization activity; acyl-CoA binding; carboxylic acid binding; manganese ion binding; metal ion binding; magnesium ion binding; receptor binding Biological Process: response to starvation; mitochondrion organization and biogenesis; leucine catabolic process; acyl-CoA metabolic process; ketone body biosynthetic process; ketone body metabolic process; cellular lipid metabolic process; liver development; protein tetramerization; response to nutrient Disease: 3-hydroxy-3-methylglutaryl-coa Lyase Deficiency
Research Articles on HMGCL	**1.** This efficient UPLC-MS/MS assay permits rapid and high sensitive determination of HMGCR enzyme activity, tracing potential alterations in cholesterol biosynthesis.
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