SANTA CRUZ BIOTECHNOLOGY, INC.

HMGCL (63Z): sc-100548

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

HMGCL (3-hydroxy-3-methylglutaryl-coenzyme A (CoA) lyase), also known as HMG-CoA lyase or HL, is a mitochondrial matrix protein that belongs to the HMG-CoA lyase family of proteins. Expressed in liver, lymphoblasts and fibroblasts, HMGCL exists as a homodimer and participates in leucine catabolism and ketogenesis, the hepatic synthesis of ketone bodies that, during fasting, provide a major source of energy for heart, brain and kidney. More specifically, HMGCL catalyzes the final step of these processes, the cleavage of 3-hydroxy-3-methylglutaryl-CoA to acetoacetic acid and acetyl-CoA. Mutations in the gene encoding HMGCL can lead to HMG-CoA lyase deficiency (also known as HL deficiency or hydroxymethylglutaricaciduria), a metabolic disease that, if left untreated, results in hypoglycemia and coma.

REFERENCES

1. Wang, S., et al. 1993. 3-hydroxy-3-methylglutaryl-coenzyme A lyase (HL): cloning and characterization of a mouse liver HL cDNA and subchromosomal mapping of the human and mouse HL genes. Mamm. Genome 4: 382-387.
2. Wang, S.P., et al. 1996. 3-hydroxy-3-methylglutaryl-CoA lyase (HL): mouse and human HL gene (HMGCL) cloning and detection of large gene deletions in two unrelated HL-deficient patients. Genomics 33: 99-104.
3. Funghini, S., et al. 2001. 3-hydroxy-3-methylglutaric aciduria in an Italian patient is caused by a new nonsense mutation in the HMGCL gene. Mol. Genet. Metab. 73: 268-275.
4. Kim, S., et al. 2004. Hepatic gene expression profiles in a long-term high-fat diet-induced obesity mouse model. Gene 340: 99-109.
5. Cardoso, M.L., et al. 2004. The E37X is a common HMGCL mutation in Portuguese patients with 3-hydroxy-3-methylglutaric-CoA lyase deficiency. Mol. Genet. Metab. 82: 334-338.
6. Al-Sayed, M., et al. 2006. Mutations underlying 3-hydroxy-3-methylglutaryl-CoA lyase deficiency in the Saudi population. BMC Med. Genet. 7: 86.
7. Alsmadi, O., et al. 2006. LCGreen I-based real-time PCR assays for detecting common ASL and HMGCL variants. Clin. Chem. 52: 1439-1440.
8. Pié, J., et al. 2007. Molecular genetics of HMG-CoA lyase deficiency. Mol. Genet. Metab. 92: 198-209.
9. Wang, X., et al. 2007. Manipulation of thyroid status and/or GH injection alters hepatic gene expression in the juvenile chicken. Cytogenet. Genome Res. 117: 174-188.

CHROMOSOMAL LOCATION

Genetic locus: HMGCL (human) mapping to 1p36.11.

SOURCE

HMGCL (63Z) is a mouse monoclonal antibody raised against recombinant HMGCL of human origin.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

HMGCL (63Z) is recommended for detection of HMGCL of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for HMGCL siRNA (h): sc-78794, HMGCL shRNA Plasmid (h): sc-78794-SH and HMGCL shRNA (h) Lentiviral Particles: sc-78794-V.

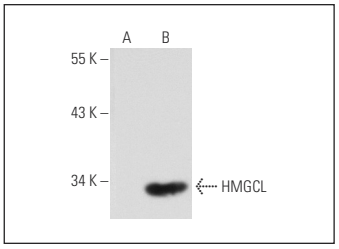
Molecular Weight of HMGCL: 31 kDa.

Positive Controls: HMGCL (h): 293 Lysate: sc-111008, A-431 whole cell lysate: sc-2201 or Hep G2 cell lysate: sc-2227.

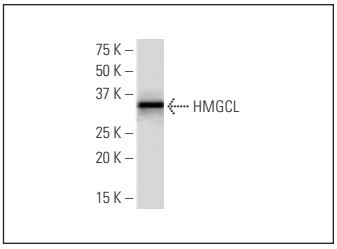
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



HMGCL (63Z): sc-100548. Western blot analysis of HMGCL expression in non-transfected: sc-110760 (**A**) and human HMGCL transfected: sc-111008 (**B**) 293 whole cell lysates.



HMGCL (63Z): sc-100548. Western blot analysis of HMGCL expression in A-431 whole cell lysate.

SELECT PRODUCT CITATIONS

1. Son, S.M., et al. 2020. Leucine regulates autophagy via acetylation of the mTORC1 component raptor. Nat. Commun. 11: 3148.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.