

# HMGCL (Y-21): sc-133661

## 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

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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.
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## CHROMOSOMAL LOCATION

Genetic locus: HMGCL (human) mapping to 1p36.11; Hmgcl (mouse) mapping to 4 D3.

## STORAGE

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

## RESEARCH USE

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

## SOURCE

HMGCL (Y-21) is an affinity purified rabbit polyclonal antibody raised against synthetic HMGCL peptide of human origin.

## PRODUCT

Each vial contains 50 µg IgG in 500 µl PBS with < 0.1% sodium azide, 0.1% gelatin and < 0.02% sucrose.

## APPLICATIONS

HMGCL (Y-21) is recommended for detection of HMGCL of mouse, rat and 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 siRNA (m): sc-146051, HMGCL shRNA Plasmid (h): sc-78794-SH, HMGCL shRNA Plasmid (m): sc-146051-SH, HMGCL shRNA (h) Lentiviral Particles: sc-78794-V and HMGCL shRNA (m) Lentiviral Particles: sc-146051-V.

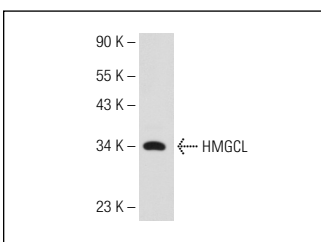
Molecular Weight of HMGCL: 31 kDa.

Positive Controls: A-431 whole cell lysate: sc-2201 or Hep G2 cell lysate: sc-2227.

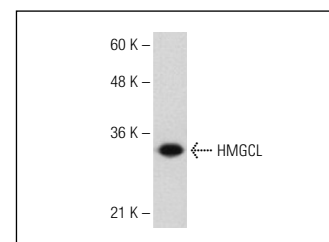
## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 (Y-21): sc-133661. Western blot analysis of HMGCL expression in A-431 whole cell lysate.



HMGCL (Y-21): sc-133661. Western blot analysis of HMGCL expression in Hep G2 whole cell lysate.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.