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# Rabbit Anti-HMGCL/FITC Conjugated antibody

SL5067R-FITC

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| **Product Name:** | Anti-HMGCL/FITC |
| **Chinese Name:** | FITC标记的三羟基三甲基辅酶A裂解酶抗体 |
| **Alias:** | 3 hydroxy 3 methylglutaryl CoA lyase; 3 hydroxy 3 methylglutaryl Coenzyme A lyase; 3 hydroxymethyl 3 methylglutaryl Coenzyme A lyase (hydroxymethylglutaricaciduria); 3 hydroxymethyl 3 methylglutaryl Coenzyme A lyase; 3-hydroxy-3-methylglutarate-CoA lyase; HL; HMG CoA lyase; HMG CoA Lyase Deficiency; HMG-CoA lyase; HMGCL; HMGCL_HUMAN; Hydroxymethylglutaricaciduria; Hydroxymethylglutaryl CoA lyase; Hydroxymethylglutaryl CoA lyase mitochondrial; Hydroxymethylglutaryl-CoA lyase; mitochondrial; MS725; OTTHUMP00000044830. |
| **Organism Species:** | Rabbit |
| **Clonality:** | Polyclonal |
| **React Species:** | Human,Mouse,Rat,Chicken,Dog,Horse,Rabbit, |
| **Applications:** | IF=1:50-200<br>not yet tested in other applications.<br>optimal dilutions/concentrations should be determined by the end user. |
| **Molecular weight:** | 32kDa |
| **Form:** | Lyophilized or Liquid |
| **Concentration:** | 1mg/ml |
| **immunogen:** | KLH conjugated synthetic peptide derived from human HMGCL |
| **Lsotype:** | IgG |
| **Purification:** | affinity purified by Protein A |
| **Storage Buffer:** | 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol. |
| **Storage:** | Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C. |
| **Product Detail:** | **background:**<br>Hydroxymethylglutaryl-CoA lyase (HMGCL) is found in fibroblasts, liver and lymphoblasts. It has a role in ketogenesis and leucine catabolism. Defects in HMGCL are the cause of 3-hydroxy-3-methylglutaryl-CoA lyase deficiency |

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| | (hydroxymethylglutaricaciduria), an autosomal recessive disease which can lead to hypoglycemia and coma.<br><br>**Function:**<br>Key enzyme in ketogenesis (ketone body formation). Terminal step in leucine catabolism.<br><br>**Subunit:**<br>Homodimer; disulfide-linked. Can also form homotetramers.<br><br>**Subcellular Location:**<br>Mitochondrion matrix.<br><br>**Tissue Specificity:**<br>Fibroblasts, liver and lymphoblasts.<br><br>**DISEASE:**<br>Defects in HMGCL are the cause of 3-hydroxy-3-methylglutaryl-CoA lyase deficiency (HMGCLD) [MIM:246450]; 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.<br><br>**Similarity:**<br>Belongs to the HMG-CoA lyase family.<br><br>**Database links:**<br>UniProtKB/Swiss-Prot: P35914.2<br><br>**Important Note:**<br>This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |