abcam

Product datasheet

Anti-HMGCL antibody ab97293

1 References 2 图像

概述

产品名称 Anti-HMGCL抗体

描述 兔多克隆抗体to HMGCL

宿主 Rabbit

经测试应用 适用于: WB, ICC/IF

种属反应性 与反应: Human

预测可用于: Mouse, Rat, Cow 📤

免疫原 Recombinant protein fragment corresponding to a region within amino acids 1 and 185 of Human

HMGCL (NP_000182).

阳性对照 WB: 293T, A431, H1299, HeLa, HepG2, MOLT4 and Raji cell lysates ICC/IF: HeLa cells

常规说明

The Life Science industry has been in the grips of a reproducibility crisis for a number of years.

Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

性能

形式 Liquid

存放说明 Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

存储溶液 pH: 7.00

Preservative: 0.01% Thimerosal (merthiolate)

Constituents: 1.21% Tris, 0.75% Glycine, 10% Glycerol (glycerin, glycerine)

纯**度** Immunogen affinity purified

纯**化**说明 Purified by antigen affinity chromatography.

 克隆
 多克隆

 同种型
 IgG

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The Abpromise guarantee

Abpromise™承诺保证使用ab97293于以下的经测试应用

"应用说明"部分下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB		1/500 - 1/3000. Predicted molecular weight: 34 kDa.
ICC/IF		1/100 - 1/200.

靶标

功能 Involved in the catabolism of branched amino acids such as leucine.

组织**特异性** Fibroblasts, liver and lymphoblasts.

通路 Metabolic intermediate metabolism; (S)-3-hydroxy-3-methylglutaryl-CoA degradation;

acetoacetate from (S)-3-hydroxy-3-methylglutaryl-CoA: step 1/1.

疾病相关 Defects in HMGCL are the cause of 3-hydroxy-3-methylglutaryl-CoA lyase deficiency (HMGCLD)

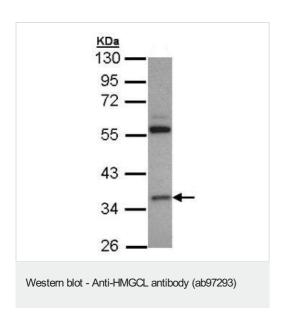
[MIM:246450]; also known as hydroxymethylglutaricaciduria or HL deficiency. An autosomal recessive disease affecting ketogenesis and L-leucine catabolism. The disease usually appears in the first year of life after a fasting period and its clinical acute symptoms include vomiting, seizures, metabolic acidosis, hypoketotic hypoglycemia and lethargy. These symptoms

sometimes progress to coma, with fatal outcome in some cases.

序列相似性 Belongs to the HMG-CoA lyase family.

细胞定位 Mitochondrion matrix.

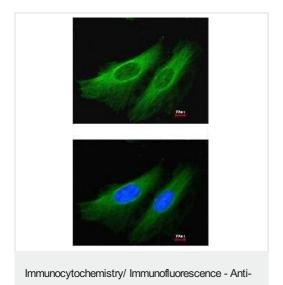
图片



Anti-HMGCL antibody (ab97293) at 1/3000 dilution + H1299 whole cell lysate at 30 μg

Predicted band size: 34 kDa

10% SDS PAGE



HMGCL antibody (ab97293)

ab97293, at a 1/200 dilution, staining HMGCL in paraformaldehyde fixed HeLa by Immunofluorescence analysis.

The lower image was merged with DNA probe.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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