abcam

Product datasheet

Recombinant Human HMGCL protein (His tag) ab226881

1 Image

Description

Product name	Recombinant Human HMGCL protein (His tag)
Purity	> 90 % SDS-PAGE. Affinity purified
Endotoxin level	< 1.000 Eu/µg
Expression system	Baculovirus infected insect cells
Accession	**P35914**
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MTLPKRVKIVEVGPRDGLQNEKNIVSTPVKIKLIDMLSEAG LSVIETTSF VSPKWVPQMGDHTEVLKGIQKFPGINYPVLTPNLKGFEAA VAAGAKEVVI FGAASELFTKKNINCSIEESFQRFDAILKAAQSANISVRGYV SCALGCPY EGKISPAKVAEVTKKFYSMGCYEISLGDTIGVGTPGIMKDM LSAVMQEVP LAALAVHCHDTYGQALANTLMALQMGVSVVDSSVAGLGG CPYAQGASGNL ATEDLVYMLEGLGIHTGVNLQKLLEAGNFICQALNRKTSSK VAQATCKLH HHHHH
Predicted molecular weight	33 kDa including tags
Amino acids	28 to 325
Tags	His tag C-Terminus
Additional sequence information	This product is the mature full length protein from aa 28 to 325. The transit peptide is not included (NP_000182).

Specifications

Our **Abpromise guarantee** covers the use of **ab226881** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	SDS-PAGE
Form	Liquid

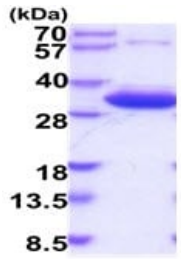
Preparation and Storage

Stability and Storage	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle. pH: 7.40 Constituents: 0.02% DTT, 20% Glycerol (glycerin, glycerine), PBS

General Info

Function	Involved in the catabolism of branched amino acids such as leucine.
Tissue specificity	Fibroblasts, liver and lymphoblasts.
Pathway	Metabolic intermediate metabolism; (S)-3-hydroxy-3-methylglutaryl-CoA degradation; acetoacetate from (S)-3-hydroxy-3-methylglutaryl-CoA: step 1/1.
Involvement in disease	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.
Sequence similarities	Belongs to the HMG-CoA lyase family.
Cellular localization	Mitochondrion matrix.

Images



SDS-PAGE - Recombinant Human HMGCL protein (His tag) (ab226881)

15% SDS-PAGE analysis of 3 µg ab226881.

MW: 28-40 kDa (SDS-PAGE under reducing conditions).

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- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit **https://www.abcam.com/abpromise** or contact our technical team.

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