abcam

Product datasheet

Recombinant Human HIBCH protein ab124585

1 Image

Description

Product name	Recombinant Human HIBCH protein
Purity	> 90 % SDS-PAGE. ab124585 is purified using conventional chromatography techniques.
Expression system	Escherichia coli
Accession	**Q6NVY1**
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MGSSHHHHHH SSGLVPRGSH MGSHMDAAEE VLLEKKGCTG VITLNRPKFL NALTLNMIRQ IYPQLKKWEQ DPETFLIIIK GAGGKAFCAG GDIRVISEAE KAKQKIAPVF FREEYMLNNA VGSCQKPYVA LIHGITMGGG VGLSVHGQFR VATEKCLFAM PETAIGLFPD VGGGYFLPRL QGKLGYFLAL TGFRLKGRDV YRAGIATHFV DSEKLAMLEE DLLALKSPSK ENIASVLENY HTESKIDRDK SFILEEHMDK INSCFSANTV EEIIENLQQD GSSFALEQLK VINKMSPTSL KITLRQLMEG SSKTLQEVLT MEYRLSQACM RGHDFHEGVR AVLIDKDQSP KWKPADLKEV TEEDLNNHFK SLGSSDLKF
Predicted molecular weight	42 kDa including tags
Amino acids	33 to 386
Tags	His tag N-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab124585** 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 Mass Spectrometry
Mass spectrometry	MALDI-TOF

Form	Liquid

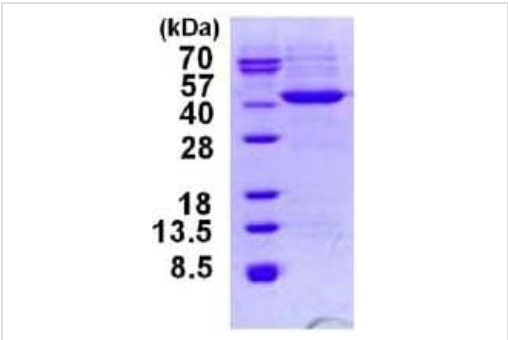
Preparation and Storage

Stability and Storage	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles. pH: 8.00 Constituents: 0.02% DTT, 0.32% Tris HCl, 10% Glycerol (glycerin, glycerine), 1.17% Sodium chloride

General Info

Function	Hydrolyzes 3-hydroxyisobutyryl-CoA (HIBYL-CoA), a saline catabolite. Has high activity toward isobutyryl-CoA. Could be an isobutyryl-CoA dehydrogenase that functions in valine catabolism. Also hydrolyzes 3-hydroxypropanoyl-CoA.
Tissue specificity	Highly expressed in liver and kidney, also detected in heart, muscle and brain (at protein level). Not detected in lung.
Pathway	Amino-acid degradation; L-valine degradation.
Involvement in disease	Defects in HIBCH are the cause of HIBCH deficiency (HIBCHD) [MIM:250620]; also known as deficiency of beta-hydroxyisobutyryl CoA deacylase or methacrylic aciduria. The enzyme defect results in accumulation of methacrylyl-CoA, a highly reactive compound, which readily undergoes addition reactions with free sulfhydryl groups. Affected individuals showed delayed development of motor skills, hypotonia, initial poor feeding, and a deterioration in neurological function during first stages of life.
Sequence similarities	Belongs to the enoyl-CoA hydratase/isomerase family.
Cellular localization	Mitochondrion.

Images



SDS-PAGE - Recombinant Human HIBCH protein (ab124585)

15% SDS-PAGE showing ab124585 at approximately 42.1kDa (3µg).

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- Replacement or refund for products not performing as stated on the datasheet
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- 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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