abcam

Product datasheet

Recombinant Human HIBCH protein ab124585

画像数 1

製品の詳細

製品名	Recombinant Human HIBCH protein
精製度	> 90 % SDS-PAGE. ab124585 is purified using conventional chromatography techniques.
発現系	Escherichia coli
アクセッション番号	Q6NVY1
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	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
予測される分子量	42 kDa including tags
領域	33 to 386
タグ	His tag N-Terminus

特性

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.

アプリケーション	SDS-PAGE
	Mass Spectrometry

質量分析	MALDI-TOF
製品の状態	Liquid

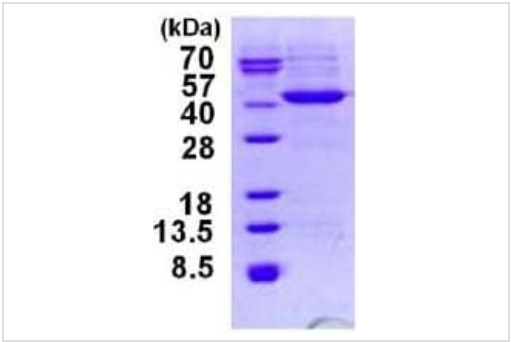
前処理および保存

保存方法および安定性	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

関連情報

機能	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.
組織特異性	Highly expressed in liver and kidney, also detected in heart, muscle and brain (at protein level). Not detected in lung.
パスウェイ	Amino-acid degradation; L-valine degradation.
関連疾患	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.
配列類似性	Belongs to the enoyl-CoA hydratase/isomerase family.
細胞内局在	Mitochondrion.

画像



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

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