abcam

Product datasheet

Recombinant Human HMBS/PBGD protein ab123176

1 图像

描述

产品名称	重组人HMBS/PBGD蛋白
纯度	> 95 % SDS-PAGE. ab123176 is purified using conventional chromatography techniques.
表达系统	Escherichia coli
Accession	P08397
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MGSSHHHHHH SSGLVPRGSH MGSHMSGNGN AAATAEENSP KMRVIRVGTR KSQLARIQTD SVVATLKASY PGLQFEIIAM STTGDKILDT ALSKIGEKSL FTKELEHALE KNEVDLVVHS LKDLPTVLPP GFTIGAICKR ENPHDAVVFH PKFVGKTLET LPEKSVVGTS SLRRAAQLQR KFPHLEFRSI RGNLNTRLRK LDEQQEFSAI ILATAGLQRM GWHNRVGQIL HPEECMYAVG QGALGVEVRA KDQDILDLVG VLHDPETLLR CIAERAFLRH LEGGCSVPVA VHTAMKDGQL YLTGGVWSLD GSDSIQETMQ ATIHVPAQHE DGPEDDPQLV GITARNIPRG PQLAAQNLGI SLANLLLSKG AKNILDVARQ LNDAH
预测分子量	42 kDa including tags
氨基酸	1 to 361
标签	His tag N-Terminus

技术指标

Our **Abpromise guarantee** covers the use of **ab123176** in the following tested applications.

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

应用	Mass Spectrometry
	SDS-PAGE

质谱法	MALDI-TOF
形式	Liquid
补充说明	This product was previously labelled as HMBS

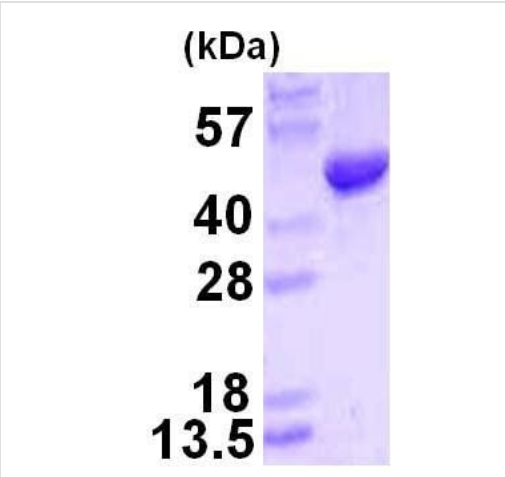
制备和贮存

稳定性和存储	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: 8.00 Constituents: 0.02% DTT, 0.32% Tris HCl, 10% Glycerol (glycerin, glycerine), 0.58% Sodium chloride

常规信息

功能	Tetrapolymerization of the monopyrrole PBG into the hydroxymethylbilane pre-uroporphyrinogen in several discrete steps.
组织特异性	Isoform 1 is ubiquitously expressed. Isoform 2 is found only in erythroid cells.
通路	Porphyrin metabolism; protoporphyrin-IX biosynthesis; coproporphyrinogen-III from 5-aminolevulinate: step 2/4.
疾病相关	Defects in HMBS are the cause of acute intermittent porphyria (AIP) [MIM:176000]. AIP is a form of porphyria. Porphyrias are inherited defects in the biosynthesis of heme, resulting in the accumulation and increased excretion of porphyrins or porphyrin precursors. They are classified as erythropoietic or hepatic, depending on whether the enzyme deficiency occurs in red blood cells or in the liver. AIP is an autosomal dominant form of hepatic porphyria characterized by acute attacks of neurological dysfunctions with abdominal pain, hypertension, tachycardia, and peripheral neuropathy. Most attacks are precipitated by drugs, alcohol, caloric deprivation, infections, or endocrine factors.
序列相似性	Belongs to the HMBS family.
细胞定位	Cytoplasm.

图片



SDS-PAGE - Recombinant Human HMBS/PBGD protein (ab123176)

15% SDS-PAGE analysis of ab123176 (3ug)

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

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