abcam

Product datasheet

Recombinant Human ALAS2/ASB protein ab79941

1 图像

描述

产品名称	重组人ALAS2/ASB蛋白
纯度	> 90 % SDS-PAGE.
表达系统	Escherichia coli
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Human
氨基酸	136 to 553
标签	His tag N-Terminus

技术指标

Our **Abpromise guarantee** covers the use of **ab79941** in the following tested applications.

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

应用	SDS-PAGE
形式	Liquid
补充说明	This product was previously labelled as ALAS2

制备和贮存

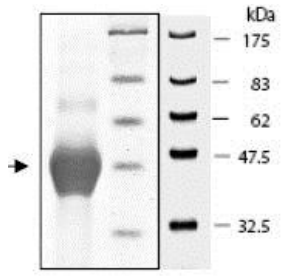
稳定性和存储	Shipped on Dry Ice. Upon delivery aliquot. Store at -80°C. Avoid freeze / thaw cycle. pH: 8.00 Constituents: 0.0462% (R*,R*)-1,4-Dimercaptobutan-2,3-diol, 0.395% Tris HCl, 0.05% Tween, 20% Glycerol (glycerin, glycerine), 0.58% Sodium chloride, 0.00053% PLP

常规信息

组织特异性	Erythroid specific.

通路	Porphyrin metabolism; protoporphyrin-IX biosynthesis; 5-aminolevulinate from glycine: step 1/1.
疾病相关	Defects in ALAS2 are a cause of anemia sideroblastic X-linked (XLSA) [MIM:300751]. Sideroblastic anemia is characterized by anemia of varying severity, hypochromic peripheral erythrocytes, systemic iron overload secondary to chronic ineffective erythropoiesis, and the presence of bone marrow ringed sideroblasts. Sideroblasts are characterized by iron-loaded mitochondria clustered around the nucleus. XLSA shows a variable hematologic response to pharmacologic doses of pyridoxine. Defects in ALAS2 are the cause of erythropoietic protoporphyria X-linked dominant (XLDPT) [MIM:300752]. 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. XLDPT is a form of porphyria characterized biochemically by a high proportion of zinc-protoporphyrin in erythrocytes, in which a mismatch between protoporphyrin production and the heme requirement of differentiating erythroid cells leads to overproduction of protoporphyrin in amounts sufficient to cause photosensitivity and liver disease. Note=Gain of function mutations in ALS2 are responsible for XLDPT, but they can also be a possible aggravating factor in congenital erythropoietic porphyria and other erythropoietic disorders caused by mutations in other genes (PubMed:21309041).
序列相似性	Belongs to the class-II pyridoxal-phosphate-dependent aminotransferase family.
细胞定位	Mitochondrion matrix.

图片



SDS-PAGE - Recombinant Human ALAS2/ASB protein (ab79941)

105 SDS-PAGE showing ab79941 at approximately 46kDa (8µg).

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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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