

Product datasheet

Recombinant Human ACADL protein ab113579

1 图像

概述

产品名称	Recombinant人ACADL protein
蛋白长度	Full length protein

描述

性质	Recombinant
来源	Escherichia coli

氨基酸序列

Accession [P28330](#)

种属 Human

序列
 MGSSHHHHHSSGLVPRGSHMGGEERLETPSAKKLTDIGIRRIFSPEHDI
 FRKSVRKFFQEEVIPHHSEWEKAGEVSREVWEKAGKQGLLGVNIAEHLGG
 IGGDLYSAAIVWEEQAYSNCSPGFSIHSGIVMSYITNHGSEEQIKHFIP
 QMTAGKCIGAIAMTEPGAGSDLQGIKTNAKKDGSDWILNGSKVFIISNGSL
 SDVVIVVAVTNHEAPSPAHGISLFLVENGMKGFIKGRKLHKMGLKAQDTA
 ELFFEDIRLPASALLGEENKGFYYIMKELPQERLLIADVAISASEFMFEE
 TRNYVKQRKAFGKTVAHLQTVQHKLAELKTHICVTRAFVDNCLQLHEAKR
 LDSATACMAKYWASELQNSVAYDCVQLHGGWGYMWEYPIAKAYVDARVQP
 IYGGTNEIMKELIAREIVFDK

分子量 47 kDa including tags

氨基酸 31 to 430

标签 His tag N-Terminus

技术指标

Our [Abpromise guarantee](#) covers the use of **ab113579** 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
	Mass Spectrometry
质谱法	MALDI-TOF
纯度	> 85 % SDS-PAGE.

> 85 % by SDS - PAGE. ab113579 is purified using conventional chromatography techniques.

形式 Liquid

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: 0.32% Tris HCl, 10% Glycerol, 0.02% DTT, 0.88% Sodium chloride

常规信息

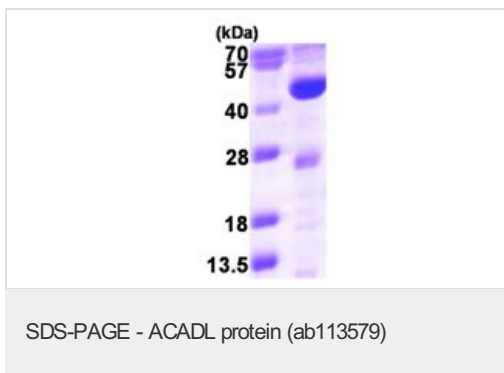
通路 Lipid metabolism; mitochondrial fatty acid beta-oxidation.

疾病相关 Defects in ACADL are a cause of acyl-CoA dehydrogenase very long-chain deficiency (ACADVLD) [MIM:201475]. An inborn error of mitochondrial fatty acid beta-oxidation which leads to impaired long-chain fatty acid beta-oxidation. It is clinically heterogeneous, with three major phenotypes: a severe childhood form characterized by early onset, high mortality and high incidence of cardiomyopathy; a milder childhood form with later onset, characterized by hypoketotic hypoglycemia, low mortality and rare cardiomyopathy; an adult form, with isolated skeletal muscle involvement, rhabdomyolysis and myoglobinuria, usually triggered by exercise or fasting.

序列相似性 Belongs to the acyl-CoA dehydrogenase family.

细胞定位 Mitochondrion matrix.

图片



15% SDS-PAGE showing ab113579 (3µg).

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