abcam

Product datasheet

Recombinant Human Caveolin-3 protein ab114265

1 References 1 图像

描述

产品名称	重组人Caveolin-3蛋白
表达系统	Wheat germ
Accession	**P56539**
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Human
序列	MMAEEHTDLEAQIVKDIHCKEIDLVNRDPKNINEDIVKVDFE DVIAEPVG TYSFDGVWKVSYTTFTVSKYWCYRLLSTLLGVP
预测分子量	35 kDa including tags
氨基酸	1 to 83

技术指标

Our **Abpromise guarantee** covers the use of **ab114265** in the following tested applications.

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

应用	ELISA
	SDS-PAGE
	Western blot
形式	Liquid
补充说明	

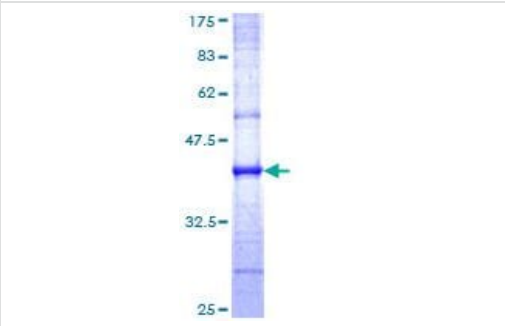
制备和贮存

稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles.
	pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

常规信息

功能	May act as a scaffolding protein within caveolar membranes. Interacts directly with G-protein alpha subunits and can functionally regulate their activity. May also regulate voltage-gated potassium channels. Plays a role in the sarcolemma repair mechanism of both skeletal muscle and cardiomyocytes that permits rapid resealing of membranes disrupted by mechanical stress.
组织特异性	Expressed predominantly in muscle.
疾病相关	Defects in CAV3 are the cause of limb-girdle muscular dystrophy type 1C (LGMD1C) [MIM:607801]. LGMD1C is a myopathy characterized by calf hypertrophy and mild to moderate proximal muscle weakness. LGMD1C inheritance can be autosomal dominant or recessive. Defects in CAV3 are a cause of hyperCKmia (HYPCK) [MIM:123320]. It is a disease characterized by persistent elevated levels of serum creatine kinase without muscle weakness. Defects in CAV3 are a cause of rippling muscle disease (RMD) [MIM:606072]. RMD is a rare disorder characterized by mechanically triggered contractions of skeletal muscle. In RMD, mechanical stimulation leads to electrically silent muscle contractions that spread to neighboring fibers that cause visible ripples to move over the muscle. Defects in CAV3 are a cause of cardiomyopathy familial hypertrophic (CMH) [MIM:192600]; also designated FHC or HCM. Familial hypertrophic cardiomyopathy is a hereditary heart disorder characterized by ventricular hypertrophy, which is usually asymmetric and often involves the interventricular septum. The symptoms include dyspnea, syncope, collapse, palpitations, and chest pain. They can be readily provoked by exercise. The disorder has inter- and intrafamilial variability ranging from benign to malignant forms with high risk of cardiac failure and sudden cardiac death. Defects in CAV3 are the cause of long QT syndrome type 9 (LQT9) [MIM:611818]. Long QT syndromes are heart disorders characterized by a prolonged QT interval on the ECG and polymorphic ventricular arrhythmias. They cause syncope and sudden death in response to excercise or emotional stress. They can present with a sentinel event of sudden cardiac death in infancy. Defects in CAV3 can be a cause of sudden infant death syndrome (SIDS) [MIM:272120]. SIDS is the sudden death of an infant younger than 1 year that remains unexplained after a thorough case investigation, including performance of a complete autopsy, examination of the death scene, and review of clinical history. Pathophysiologic mechanisms for SIDS may include respiratory dysfunction, cardiac dysrhythmias, cardiorespiratory instability, and inborn errors of metabolism, but definitive pathogenic mechanisms precipitating an infant sudden death remain elusive. Long QT syndromes-associated mutations can be responsible for some SIDS cases.
序列相似性	Belongs to the caveolin family.
细胞定位	Golgi apparatus membrane. Cell membrane. Membrane > caveola. Potential hairpin-like structure in the membrane. Membrane protein of caveolae.

图片



SDS-PAGE - Recombinant Human Caveolin-3 protein (ab114265)

ab114265 analysed on a 12.5% SDS-PAGE gel stained with Coomassie Blue.

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