abcam

Product datasheet

Recombinant Human Dystrophin protein ab114197

1 图像

概述

产品名称	重组人Dystrophin蛋白
蛋白长度	Full length protein

描述

性质	Recombinant
来源	Wheat germ

氨基酸序列

种属	Human
序列	MREQLKGHETQTTCWDHPKMTELYQSLADLNNVRFSAYRTAMKLRRLQKA LCLDLLSLSAACDALDQHNLKQNDQPMDILQIINCLTTIYDRLEQEHNNL VNVPLCVDMCLNWLLNVYDTGRTGRIRVLSFKTGIISLCKAHLEDKYRYL FKQVASSTGFCDQRRLGLLLHDSIQIPRQLGEVASFGGSNIEPSVRSCFQ FANNKPEIEAALFLDWMRLEPQSMVWLPVLHRVAAAETAKHQAKCNICKE CPIIGFRYRSLKHFNYDICQSCFFSGRVAKGHKMHYPMVEYCTPTTSGED VRDFAKVLKNKFRTKRYFAKHPRMGYLPVQTVLEGDNMETPVTLINFWPV DSAPASSPQLSHDDTHSRIEHYASRLAEMENSNGSYLNDSISPNESIDDE HLLIQHYCQSLNQDSPLSQPRSPAQILISLESEERGELERILADLEEENR NLQAEYDRLKQQHEHKGLSPLPSPPEMMPTSPQSPRDAELIAEAKLLRQH KGRLEARMQILEDHNKQLESQLHRLRQLLEQPQAEAKVNGTTVSSPSTSL QRSDSSQPMLLRVVGSQTSDSMGEEDLLSPPQDTSTGLEEVMEQLNNSFP SSRGHNVGSLFHMADDLGRAMESLVSVMTDEEGAE
分子量	96 kDa including tags
氨基酸	1 to 635

技术指标

Our Abpromise guarantee covers the use of **ab114197** 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	Protein concentration is above or equal to 0.05 µg/µl. Best use within three months from the date of receipt of this protein.

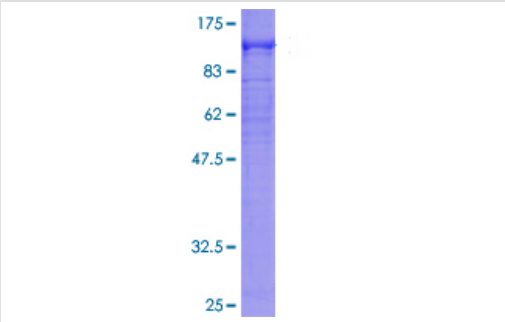
制备和贮存

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	Anchors the extracellular matrix to the cytoskeleton via F-actin. Ligand for dystroglycan. Component of the dystrophin-associated glycoprotein complex which accumulates at the neuromuscular junction (NMJ) and at a variety of synapses in the peripheral and central nervous systems and has a structural function in stabilizing the sarcolemma. Also implicated in signaling events and synaptic transmission.
组织特异性	Expressed in muscle fibers accumulating in the costameres of myoplasm at the sarcolemma. Expressed in brain, muscle, kidney, lung and testis. Isoform 5 is expressed in heart, brain, liver, testis and hepatoma cells. Most tissues contain transcripts of multiple isoforms, however only isoform 5 is detected in heart and liver.
疾病相关	Defects in DMD are the cause of Duchenne muscular dystrophy (DMD) [MIM:310200]. DMD is the most common form of muscular dystrophy; a sex-linked recessive disorder. It typically presents in boys aged 3 to 7 year as proximal muscle weakness causing waddling gait, toe-walking, lordosis, frequent falls, and difficulty in standing up and climbing up stairs. The pelvic girdle is affected first, then the shoulder girdle. Progression is steady and most patients are confined to a wheelchair by age of 10 or 12. Flexion contractures and scoliosis ultimately occur. About 50% of patients have a lower IQ than their genetic expectations would suggest. There is no treatment. Defects in DMD are the cause of Becker muscular dystrophy (BMD) [MIM:300376]. BMD resembles DMD in hereditary and clinical features but is later in onset and more benign. Defects in DMD are a cause of cardiomyopathy dilated X-linked type 3B (CMD3B) [MIM:302045]; also known as X-linked dilated cardiomyopathy (XLCM). Dilated cardiomyopathy is a disorder characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia. Patients are at risk of premature death.
序列相似性	Contains 2 CH (calponin-homology) domains. Contains 22 spectrin repeats. Contains 1 WW domain. Contains 1 ZZ-type zinc finger.
细胞定位	Cell membrane > sarcolemma. Cytoplasm > cytoskeleton.

图片



SDS-PAGE - Dystrophin protein (ab114197)

12.5% SDS-PAGE showing ab114197 at approximately 95.96kDa stained with Coomassie Blue.

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