abcam

Product datasheet

Recombinant Human Dystrophin protein (Tagged) ab114197

1 图像

描述

产品名称	重组人Dystrophin蛋白(Tagged)
表达系统	Wheat germ
Accession	P11532
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Human
序列	MREQLKGHETQTTCWDHPKMTELYQSLADLNNVRFSAYRTAM KLRRLQKA LCLDLLSLSAACDALDQHNLKQNDQPMDILQIINCLTTIYDR LEQEHNNL VNVPLCVDMCLNWLLNVYDTGRTGRIRVLSFKTGIISLCKAH LEDKYRYL FKQVASSTGFCDQRRLGLLLHDSIQIPRQLGEVASFGGSNIE PSVRSCFQ FANNKPEIEAALFLDWMRLEPQSMVWLPVLHRVAAAETAKHQ AKCNICKE CPIIGFRYRSLKHFNYDICQSCFFSGRVAKGHKMHYPMVEYC TPTTSGED VRDFAKVLKNKFRTKRYFAKHPRMGYLPVQTVLEGDNMETPV TLINFWPV DSAPASSPQLSHDDTHSRIEHYASRLAEMENSNGSYLNDSIS PNESIDDE HLLIQHYCQSLNQDSPLSQPRSPAQILISLESEERGELERIL ADLEEENR NLQAEYDRLKQQHEHKGLSPLPSPPEMMPTSPQSPRDAELIA EAKLLRQH KGRLEARMQILEDHNKQLESQLHRLRQLLEQPQAEAKVNGTT VSSPSTSL QRSDSSQPMLLRVVGSQTSDSMGEEDLLSPPQDTSTGLEEVM EQLNNSFP SSRGHNVGSLFHMADDLGRAMESLVSVMTDEEGAE
预测分子量	96 kDa including tags
氨基酸	3076 to 3674

标签	GST tag N-Terminus

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

制备和贮存

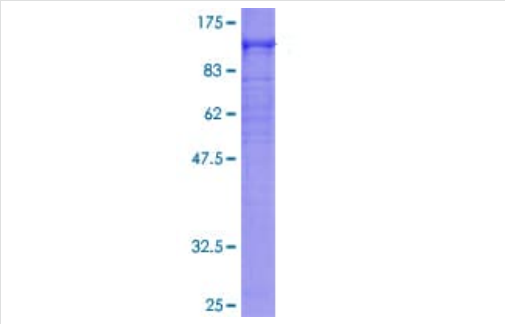
稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

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 - Recombinant Human Dystrophin protein (ab114197)

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

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