abcam

Product datasheet

Recombinant Human Dystrophin protein (Tagged) ab114197

画像数 1

製品の詳細

製品名	Recombinant Human Dystrophin protein (Tagged)
発現系	Wheat germ
アクセッション番号	P11532
タンパク質長	Protein fragment
Animal free	No
由来	Recombinant
生物種	Human
配列	MREQLKGHETQTTCWDHPKMTELYQSLADLNNVRFSAYRTAMKLRRLQKA LCLDLLSLSAACDALDQHNLKQNDQPMDILQIINCLTTIYDRLEQEHNNL VNVPLCVDMCLNWLLNVYDTGRTGRIRVLSFKTGIISLCKAHLEDKYRYL FKQVASSTGFCDQRRLGLLLHDSIQIPRQLGEVASFGGSNIEPSVRSCFQ FANNKPEIEAALFLDWMRLEPQSMVWLPVLHRVAAAETAKHQAKCNICKE CPIIGFRYRSLKHFNYDICQSCFFSGRVAKGHKMHYPMVEYCTPTTSGED VRDFAKVLKNKFRTKRYFAKHPRMGYLPVQTVLEGDNMETPVTLINFWPV DSAPASSPQLSHDDTHSRIEHYASRLAEMENSNGSYLNDSISPNESIDDE HLLIQHYCQSLNQDSPLSQPRSPAQILISLESEERGELERILADLEEENR NLQAEYDRLKQQHEHKGLSPLPSPPEMMPTSPQSPRDAELIAEAKLLRQH KGRLEARMQILEDHNKQLESQLHRLRQLLEQPQAEAKVNGTTVSSPSTSL QRSDSSQPMLLRVVGSQTSDSMGEEDLLSPPQDTSTGLEEVMEQLNNSFP SSRGHNVGSLFHMADDLGRAMESLVSVMTDEEGAE
予測される分子量	96 kDa including tags
領域	3076 to 3674

タグ	GST tag N-Terminus

特性

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.

アプリケーション	ELISA SDS-PAGE Western blot
製品の状態	Liquid

前処理および保存

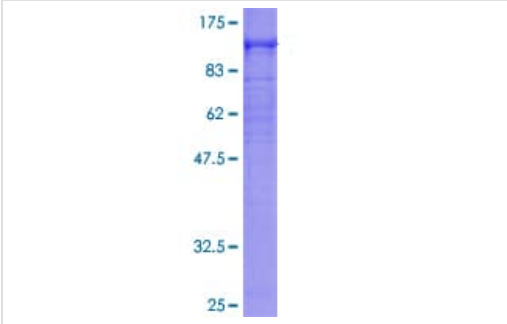
保存方法および安定性	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

関連情報

機能	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.
組織特異性	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.
関連疾患	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.
配列類似性	Contains 2 CH (calponin-homology) domains. Contains 22 spectrin repeats. Contains 1 WW domain. Contains 1 ZZ-type zinc finger.

細胞内局在	Cell membrane > sarcolemma. Cytoplasm > cytoskeleton.

画像



SDS-PAGE - Recombinant Human Dystrophin protein (ab114197)

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

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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