abcam

Product datasheet

Recombinant Human Titin protein (Tagged) ab237002

画像数 1

製品の詳細

製品名	Recombinant Human Titin protein (Tagged)
精製度	> 85 % SDS-PAGE.
発現系	Escherichia coli
アクセッション番号	Q8WZ42
タンパク質長	Protein fragment
Animal free	No
由来	Recombinant
生物種	Human
配列	RCEEGKDNWIRCNMKLVPELTYKVTGLEKGNKYLYRVSAENK AGVSDPSE ILGPLTADDAFVEPTMDLSAFKDGLEVIVPNPITILVPSTGY PRPTATWC FGDKVLETGDRVKMKTLSAYAELVISPSERSDKGIYTLKLEN RVKTISGE IDVNVIARPSAPKELKFGDITKDSVHLTWEPPDDDGGSPLTG YVVEKREV SRKTWTKVMDFVTDLEFTVPDLVQGKEYLFKVCARNKCGPGE PAYVDEPV NMSTPATVPDPPENVKWRDRTANSIFLTWDPPKNDGG
予測される分子量	37 kDa including tags
領域	14257 to 14543
配列の追加情報	N-terminal 10xHis-tagged and C-terminal Myc-tagged.

特性

Our **Abpromise guarantee** covers the use of **ab237002** 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	SDS-PAGE
製品の状態	Liquid

前処理および保存

前処理および保存

保存方法および安定性

Shipped at 4°C. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

pH: 7.2
Constituents: Tris buffer, 50% Glycerol (glycerin, glycerine)

関連情報

機能

Key component in the assembly and functioning of vertebrate striated muscles. By providing connections at the level of individual microfilaments, it contributes to the fine balance of forces between the two halves of the sarcomere. The size and extensibility of the cross-links are the main determinants of sarcomere extensibility properties of muscle. In non-muscle cells, seems to play a role in chromosome condensation and chromosome segregation during mitosis. Might link the lamina network to chromatin or nuclear actin, or both during interphase.

組織特異性

Isoform 3, isoform 7 and isoform 8 are expressed in cardiac muscle. Isoform 4 is expressed in vertebrate skeletal muscle. Isoform 6 is expressed in cardiac tissues.

関連疾患

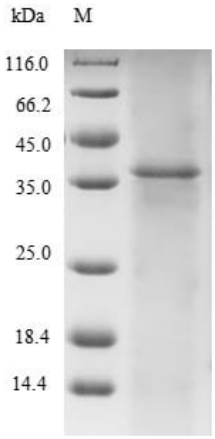
Defects in TTN are the cause of hereditary myopathy with early respiratory failure (HMERF) [MIM:603689]; also known as Edstrom myopathy. HMERF is an autosomal dominant, adult-onset myopathy with early respiratory muscle involvement.
Defects in TTN are the cause of familial hypertrophic cardiomyopathy type 9 (CMH9) [MIM:613765]. 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 TTN are the cause of cardiomyopathy dilated type 1G (CMD1G) [MIM:604145]. 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.
Defects in TTN are the cause of tardive tibial muscular dystrophy (TMD) [MIM:600334]; also known as Udd myopathy. TMD is an autosomal dominant, late-onset distal myopathy. Muscle weakness and atrophy are usually confined to the anterior compartment of the lower leg, in particular the tibialis anterior muscle. Clinical symptoms usually occur at age 35-45 years or much later.
Defects in TTN are the cause of limb-girdle muscular dystrophy type 2J (LGMD2J) [MIM:608807]. LGMD2J is an autosomal recessive degenerative myopathy characterized by progressive weakness of the pelvic and shoulder girdle muscles. Severe disability is observed within 20 years of onset.
Defects in TTN are the cause of early-onset myopathy with fatal cardiomyopathy (EOMFC) [MIM:611705]. Early-onset myopathies are inherited muscle disorders that manifest typically from birth or infancy with hypotonia, muscle weakness, and delayed motor development. EOMFC is a titinopathy that, in contrast with the previously described examples, involves both heart and skeletal muscle, has a congenital onset, and is purely recessive. This phenotype is due to homozygous out-of-frame TTN deletions, which lead to a total absence of titin's C-terminal end from striated muscles and to secondary CAPN3 depletion.

配列類似性

Belongs to the protein kinase superfamily. CAMK Ser/Thr protein kinase family.
Contains 132 fibronectin type-III domains.
Contains 152 Ig-like (immunoglobulin-like) domains.
Contains 19 Kelch repeats.
Contains 1 protein kinase domain.
Contains 17 RCC1 repeats.

	Contains 14 TPR repeats. Contains 15 WD repeats.
ドメイン	ZIS1 and ZIS5 regions contain multiple SPXR consensus sites for ERK- and CDK-like protein kinases as well as multiple SP motifs. ZIS1 could adopt a closed conformation which would block the TCAP-binding site. The PEVK region may serve as an entropic spring of a chain of structural folds and may also be an interaction site to other myofilament proteins to form interfilament connectivity in the sarcomere.
翻訳後修飾	Autophosphorylated (By similarity). Phosphorylated upon DNA damage, probably by ATM or ATR.
細胞内局在	Cytoplasm. Nucleus.

画像



SDS-PAGE - Recombinant Human Titin protein (Tagged) (ab237002)

(Tris-Glycine gel) Discontinuous SDS-PAGE (reduced) with 5% enrichment gel and 15% separation gel of ab237002.

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