




Product datasheet

Anti-Titin antibody ab224346

1 Image

Overview

| | |
|----------------------------|---|
| Product name | Anti-Titin antibody |
| Description | Rabbit polyclonal to Titin |
| Host species | Rabbit |
| Tested applications | Suitable for: IHC-P |
| Species reactivity | Reacts with: Human Predicted to work with: Mouse  |
| Immunogen | Recombinant fragment corresponding to Human Titin aa 1000-1200. Database link: Q8WZ42  Run BLAST with  Run BLAST with |
| Positive control | IHC-P: Human heart muscle tissue. |
| General notes | <p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As</p> |

Properties

| | |
|-----------------------------|---|
| Form | Liquid |
| Storage instructions | Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle. |
| Storage buffer | pH: 7.20 Preservative: 0.02% Sodium azide Constituents: 40% Glycerol (glycerin, glycerine), PBS |
| Purity | Immunogen affinity purified |
| Clonality | Polyclonal |
| Isotype | IgG |

Applications

The Abpromise guarantee

Our **Abpromise guarantee** covers the use of ab224346 in the following tested applications.

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

| Application | Abreviews | Notes |
|-------------|-----------|---|
| IHC-P | | 1/200 - 1/500. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol. |

Target

Function

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.

Tissue specificity

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.

Involvement in disease

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.

Sequence similarities

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.

Domain

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.

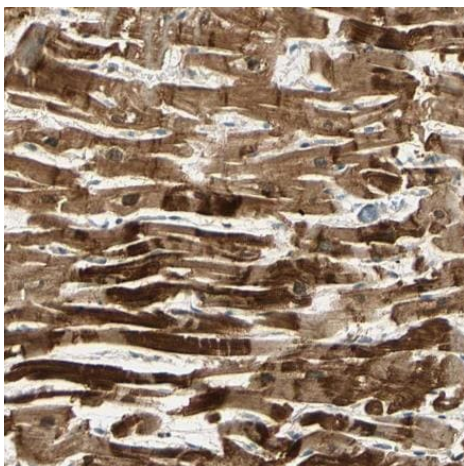
Post-translational modifications

Autophosphorylated (By similarity). Phosphorylated upon DNA damage, probably by ATM or ATR.

Cellular localization

Cytoplasm. Nucleus.

Images



Paraffin-embedded human heart muscle tissue stained for Titin using ab224346 at 1/50 in immunohistochemical analysis.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Titin antibody (ab224346)

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