


Product datasheet

Anti-Titin antibody [T11] ab7034

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Overview

<b>Product name</b>	Anti-Titin antibody [T11]
<b>Description</b>	Mouse monoclonal [T11] to Titin
<b>Host species</b>	Mouse
<b>Specificity</b>	Monoclonal Anti-Titin reacts only with skeletal and cardiac muscle, but not with smooth muscle or different non-muscle tissues and cultured cells. The antibody localizes titin (connectin) in skeletal and heart muscle of a wide variety of species from cold-blooded vertebrates to human. The product does not cross-react with nebulin.
<b>Tested applications</b>	<b>Suitable for:</b> WB, IHC-Fr, ICC/IF, Immunomicroscopy, Dot blot, Electron Microscopy
<b>Species reactivity</b>	<b>Reacts with:</b> Rat, Chicken <b>Predicted to work with:</b> Rabbit, Human, Pig, Fish, a wide range of other species  <b>Does not react with:</b> Mouse, Cow
<b>Immunogen</b>	Full length native protein (purified) (Chicken breast muscle).
<b>Positive control</b>	IHC-P: Rat soleus muscle tissue.
<b>General notes</b>	Monoclonal Anti-Titin can be used for study of the elastic filaments within sarcomeric structures. It is also useful as a differentiation marker in the separation of rhabdomyosarcomas from other muscle tumors. By indirect immunofluorescence the antibody displays a typical striated staining pattern on frozen sections of chicken skeletal and cardiac muscle tissues. Stains the region of the A-I junction by indirect immunofluorescence. It shows a decoration line 0.05 mm from the end of the A band in electron micro-graphs. In immunoblotting, using total extracts of chicken breast muscle, the antibody reacts specifically with both bands of the titin double and shows no reaction with nebulin.

Properties

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
<b>Storage buffer</b>	Preservative: 15mM Sodium Azide Constituents: Ascites
<b>Purity</b>	Ascites
<b>Primary antibody notes</b>	Monoclonal Anti-Titin can be used for study of the elastic filaments within sarcomeric structures. It is also useful as a differentiation marker in the separation of rhabdomyosarcomas from other

muscle tumors. By indirect immunofluorescence the antibody displays a typical striated staining pattern on frozen sections of chicken skeletal and cardiac muscle tissues. Stains the region of the A-I junction by indirect immunofluorescence. It shows a decoration line 0.05 mm from the end of the A band in electron micro-graphs. In immunoblotting, using total extracts of chicken breast muscle, the antibody reacts specifically with both bands of the titin double and shows no reaction with nebulin.

<b>Clonality</b>	Monoclonal
<b>Clone number</b>	T11
<b>Isotype</b>	IgG2b

## Applications

Our [Abpromise guarantee](#) covers the use of **ab7034** 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
WB		Use at an assay dependent concentration. Predicted molecular weight: 2000 kDa. Fish skeletal muscle + Toad skeletal muscle + Salamander skeletal muscle + Salamander heart muscle + Alligator skeletal muscle + Pigeon skeletal muscle + Chicken skeletal muscle + Chicken heart muscle + Chicken gizzard smooth muscle - Mouse skeletal muscle - Rat skeletal muscle + Rat heart muscle + Rat rhabdomyosarcoma + Rabbit skeletal muscle + Bovine heart muscle - Pig skeletal muscle + Human skeletal muscle +
IHC-Fr	★★★★☆	Use at an assay dependent concentration.
ICC/IF		1/1000.
Immunomicroscopy		Use at an assay dependent concentration.
Dot blot		Use at an assay dependent concentration.
Electron Microscopy		Use at an assay dependent concentration.

## 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**

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

**modifications**

ATR.

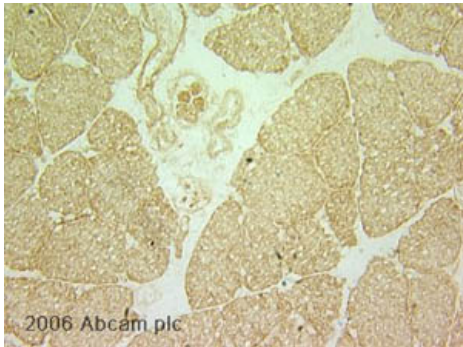
**Cellular localization**

Cytoplasm. Nucleus.

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## Images

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ab7034 at 1/100 staining rat soleus muscle tissue sections by IHC-Fr. The tissue was paraformaldehyde fixed and blocked with horse serum. A biotinylated horse anti-mouse IgG was used as the secondary.

Immunohistochemistry (Frozen sections) - Titin antibody [T11] (ab7034)

This image is courtesy of an anonymous Abreview

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