Product Name: TTN Rabbit Polyclonal Antibody
Catalog #: APRab19409

Summary

Production Name	TTN Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	IHC,ELISA
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	TTN
Alternative Names	Titin (EC 2.7.11.1;Connectin;Rhabdomyosarcoma antigen MU-RMS-40.14)
Gene ID	7273.0
SwissProt ID	Q8WZ42.Synthetic peptide from human protein at AA range: 161-210

Application

Dilution Ratio	IHC 1:50-200 ELISA 1:10000-20000
Molecular Weight	

Background

This gene encodes a large abundant protein of striated muscle. The product of this gene is divided into two regions, a N-terminal I-band and a C-terminal A-band. The I-band, which is the elastic part of the molecule, contains two regions of

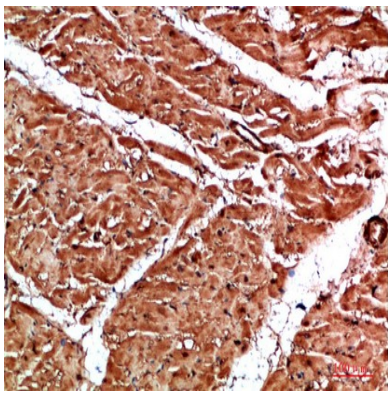
tandem immunoglobulin domains on either side of a PEVK region that is rich in proline, glutamate, valine and lysine. The A-band, which is thought to act as a protein-ruler, contains a mixture of immunoglobulin and fibronectin repeats, and possesses kinase activity. An N-terminal Z-disc region and a C-terminal M-line region bind to the Z-line and M-line of the sarcomere, respectively, so that a single titin molecule spans half the length of a sarcomere. Titin also contains binding sites for muscle associated proteins so it serves as an adhesion template for the assembly of contractile machinery in muscle cells. It has also been identified as a structural protein for chromosomes.alternative products:A number of isoforms may be produced, ranging from 27000 to 33000 residues in different striated muscle tissues, the size of the full length protein may be up to 38138 residues,catalytic activity:ATP + a protein = ADP + a phosphoprotein.,cofactor:Magnesium.,disease: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.,disease:Defects in TTN are the cause of cardiomyopathy familial hypertrophic type 9 (CMH9) [MIM:188840]. 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.,disease: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.,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.,disease: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.,disease: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.,domain: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.,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.,enzyme regulation:Full activation of the protein kinase domain requires both phosphorylation of Tyr-32341, preventing it from blocking the catalytic aspartate residue, and binding of Ca/CALM to the C-terminal regulatory tail of the molecule which results in ATP binding to the kinase.,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.,miscellaneous:In some isoforms, after the PEVK repeat region there is a long PEVK duplicated region. On account of this region, it has been very difficult to sequence the whole protein. The length of this region (ranging from 183 to 2174 residues), may be a key elastic element of titin.,online information:Titin entry,PTM:Autophosphorylated (By similarity). Phosphorylated upon DNA damage, probably by ATM or ATR.,sequence caution:Contaminating sequence. Potential poly-A sequence starting in position 553.,sequence caution:Contaminating sequence. Potential poly-A sequence starting in position 627.,similarity:Belongs to the protein kinase superfamily. CAMK Ser/Thr protein kinase family.,similarity:Contains 1 protein kinase domain.,similarity:Contains 132 fibronectin type-III domains.,similarity:Contains 14 TPR repeats.,similarity:Contains 15 WD repeats.,similarity:Contains 152 Ig-like (immunoglobulin-like) domains.,similarity:Contains 17 RCC1 repeats.,similarity:Contains 19 Kelch repeats.,subunit:Interacts with MYOM1, MYOM2, tropomyosin and myosin. Interacts with actin, primarily via the PEVK domains and with MYPN (By similarity). Interacts with FHL2, NEB, CRYAB, LMNA/lamin-A and LMNB/lamin-B. Interacts with TCAP/telethonin and/or ANK1 isoform Mu17/ank1.5, via the first two N-terminal immunoglobulin domains. Interacts with TRIM63 and TRIM55, through several domains including immunoglobulin domains 141 and 142. Interacts with ANKRD1, ANKRD2 and ANKRD23, via the region between immunoglobulin domains 77 and 78 and interacts with CAPN3, via immunoglobulin domain 79. Interacts with NBR1 through the protein kinase domain. Interacts with CALM/calmodulin. Isoform 8 interacts with OBSCN isoform 3.,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.,

Research Area

Hypertrophic cardiomyopathy (HCM);Dilated cardiomyopathy;

Image Data



Immunohistochemical analysis of paraffin-embedded human-heart, antibody was diluted at 1:200

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Note

For research use only.