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Rabbit Anti-MYBPC1 antibody

SL11034R

Product Name:	MYBPC1
Chinese Name:	肌球蛋白Binding proteinC抗体
Alias:	skeletal muscle slow isoform; slow-type; C protein, skeletal muscle slow isoform; C-protein; MYBPC1; MYBPCC; MYBPCS; Myosin binding protein C, slow type; Myosin-binding protein C; MYPC1_HUMAN; skeletal muscle C protein; Slow MyBP C; Slow MyBP-C.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	128kDa
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human MYBPC1:51-150/1141
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	MYBPC1 is a 1,141 amino acid protein that contains three fibronectin type-III domains and seven Ig-like C2-type domains. Existing as a member of the immunoglobulin superfamily, MYBPC1 functions as a thick filament-associated protein that localizes to striated muscle bands in vertebrae and is thought to modify the activity of select ATPases. Additionally, MYBPC1 may play a role in the modulation of muscle

contraction and in the overall structural integrity of the cell. The gene encoding MYBPC1 maps to human chromosome 12, which encodes over 1,100 genes and comprises approximately 4.5% of the human genome. Chromosome 12 is associated with a variety of diseases and afflictions, including hypochondrogenesis, achondrogenesis, Kniest dysplasia, Noonan syndrome and Trisomy 12p, which causes facial developmental defects and seizure disorders.

Function:
Thick filament-associated protein located in the crossbridge region of vertebrate striated muscle a bands. In vitro it binds MHC, F-actin and native thin filaments, and modifies the activity of actin-activated myosin ATPase. It may modulate muscle contraction or may play a more structural role.

Subunit:
Interacts with USP25 (isoform USP25m only); the interaction prevents proteasomal degradation of MYBPC1.

DISEASE:
Defects in MYBPC1 are the cause of arthrogryposis, distal, type 1B (DA1B) [MIM:614335]. A form of distal arthrogryposis, a disease characterized by congenital joint contractures that mainly involve two or more distal parts of the limbs, in the absence of a primary neurological or muscle disease. Distal arthrogryposis type 1 is characterized largely by camptodactyly and clubfoot. Hypoplasia and/or absence of some interphalangeal creases is common. The shoulders and hips are less frequently affected.
Note=Defects in MYBPC1 may be a cause of autosomal recessive lethal congenital contractural syndrome (LCCS), a severe, neonatally lethal form of arthrogryposis.

Similarity:
Belongs to the immunoglobulin superfamily. MyBP family.
Contains 3 fibronectin type-III domains.
Contains 7 Ig-like C2-type (immunoglobulin-like) domains.

SWISS:
Q00872

Gene ID:
4604

Database links:

Entrez Gene: 4604Human

Entrez Gene: 109272Mouse

Entrez Gene: 362867Rat

	Omim: 160794Human SwissProt: Q00872Human SwissProt: Q63518Rat Unigene: 654589Human Unigene: 147208Mouse Unigene: 9153Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.