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MYBPC1 Rabbit pAb

Catalog # AP99686

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q00872
Other Accession	Q00872
Predicted	Rat, Human, Mouse, Chicken, Dog, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	128294
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MYBPC1
Epitope Specificity	51-150/1141
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the immunoglobulin superfamily. MyBP family. Contains 3 fibronectin type-III domains. Contains 7 Ig-like C2-type (immunoglobulin-like) domains.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	4604
Other Names	CMYO16; CMYP16; LCCS4; MYBPCC; MYBPCS; MYOTREM; ssMyBP-C; 8030451F13Rik; MYPC1_HUMAN; MYBPC1; Slow MyBP-C; C-protein, skeletal muscle slow isoform; MYPC1_RAT; myosin binding protein C1; myosin-binding protein C, slow-type; slow skeletal-type muscle myosin-binding-protein C
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	MYBPC1
Synonyms	MYBPCS
Function	Thick filament-associated protein located in the crossbridge region of vertebrate striated muscle a bands. Slow skeletal protein that binds to both myosin and actin (PubMed:31025394, PubMed:31264822). In vitro, binds to native thin filaments and modifies the activity of actin-activated myosin ATPase. May modulate muscle contraction or may play a more structural role.

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