

MYBPC3 Rabbit pAb

Catalog # AP100005

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q14896
Other Accession	Q14896
Host	Rabbit
Clonality	Polyclonal
Calculated MW	140762
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MYBPC3
Epitope Specificity	161-260/1274
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.
Post-translational modifications	Substrate for phosphorylation by PKA and PKC. Reversible phosphorylation appears to modulate contraction (By similarity).
DISEASE	Defects in MYBPC3 are the cause of familial hypertrophic cardiomyopathy type 4 (CMH4) [MIM:115197]. 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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	MYBPC3 encodes the cardiac isoform of the thick-filament myosin-binding protein C. It is found in the crossbridge-bearing zone (C region) of A bands in vertebrate striated muscle. Regulatory phosphorylation of MYBPC3 by cAMP-dependent protein kinase (PKA) upon adrenergic stimulation may be linked to modulation of cardiac contraction. MYBPC3 binds F-Actin, MHC and native thin filaments, and modifies the activity of Actin-activated myosin ATPase. Mutations in the MYBPC3 gene lead mainly to truncation of the protein, which results in one cause of familial hypertrophic cardiomyopathy type 4 (CMH4), a heart disorder characterized by ventricular hypertrophy, which often involves the interventricular septum and is usually asymmetric. The MYBPC3 gene maps to chromosome 11p11.2.

Additional Information

Gene ID 4607

Other Names	CMD1MM; CMH4; FHC; LVNC10; MYBP-C; cMyBP-C; MYPC3_HUMAN; MYBPC3; Cardiac MyBP-C; C-protein, cardiac muscle isoform; MYPC3_MOUSE; MYPC_RAT; myosin binding protein C3; myosin-binding protein C, cardiac
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=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	MYBPC3
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.

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