

Human MYBPC3 Polyclonal Antibody

Antigen Affinity-Purified Anti-Human MYBPC3 Rabbit Antibody Catalog Number: GR126171

Background

The myosin-binding protein C, cardiac-type (MyBPC3 or cMyBP-C) is a protein that in humans is encoded by the MYBPC3 gene.^[1] This isoform is expressed exclusively in heart muscle during human and mouse development, [2] and is distinct from those expressed in slow skeletal muscle (MYBPC1) and fast skeletal muscle (MYBPC2). cMyBP-C is a 140.5 kDa protein composed of 1273 amino acids.^[3] cMyBP-C is a myosin-associated protein that binds at 43 nm intervals along the myosin thick filament backbone, stretching for 200 nm on either side of the M-line within the crossbridge-bearing zone (C-region) of the A band in striated muscle. The approximate stoichiometry of cMyBP-C along the thick filament is 1 per 9-10 myosin molecules, or 37 cMyBP-C molecules per thick filament. In addition to myosin, cMyBP-C also binds titin and actin. [4] The cMyBP-C isoform expressed in cardiac muscle differs from those expressed in slow and fast skeletal muscle (MYBPC1 and MYBPC2, respectively) by three features: (1) an additional immunoglobulin (Ig)-like domain on the N-terminus, (2) a linker region between the second and third Ig domains, and (3) an additional loop in the sixth Ig domain.^[5] cMyBP-C appears necessary for normal order, filament length and lattice spacing within the structure of the sarcomere. [6] cMyBP-C is not essential for sarcomere formation during embryogenesis, but is crucial for sarcomere organization and maintenance of normal cardiac function. Absence of cMyBP-C results in severe cardiac hypertrophy, increased heart-weight-tobody-weight-ratios, enlargement of ventricles, increased myofilament Ca2+ sensitivity and depressed diastolic and systolic function.^[7] Histologically, MyBPC3-targeted knock-out hearts display structural rearrangements with cardiac myocyte disarray and increased interstitial fibrosis similar to patients with hypertrophic cardiomyopathy, without obvious alterations in shape or size of single cardiac myocytes, a loss of lateral alignment of adjacent myofibrils with their Zlines misaligned.[7]

References

- 1. Gautel M, et al. (1995). The EMBO Journal. 14 (9): 1952–60.
- 2. Fougerousse F, et al. (1998). Circulation Research. **82** (1): 130–3.
- 3. Carrier L, et al. (1997). Circulation Research. **80** (3): 427–34.
- 4. Freiburg A, et al. (1996). European Journal of Biochemistry / FEBS. 235 (1–2): 317–23.
- 5. Winegrad S (1999). Circulation Research. **84** (10): 1117–26.
- 6. Colson BA, et al. (2007). Journal of Molecular Biology. **367** (1): 36–41.
- 7. Harris SP, et al. (2002). Circulation Research. **90** (5): 594–601.



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Description

Species reactivity: Human

Specificity: Detects human MYBPC3 in direct or indirect ELISAs and Western blots.

Source: Polyclonal rabbit IgG

Purification: Antigen Affinity purified

Immunogen: E. coli derived recombinant human MYBPC3, Pro2-Ser175, and Accession #

NP 000247.2.

Endotoxin Level: <0.10 EU per 1 μg of the antibody by the LAL method.

Formulation: lyophilized from a solution containing PBS and trehalose (100 µg/ml).

Application

Reconstitution: reconstitute at 0.2 mg/ml in sterile PBS

 $\frac{Recommended\ concentration}{We stern\ blot:} > 0.1\ \mu g/ml$

Immunocytochemistry: 5-15 µg/ml

ELISA: 0.2-0.6 μg/ml

Stability & Storage

Use a manual defrost freezer and avoid repeated freeze-thaw cycles.

- 12 months at -20°C as supplied.
- 1 month after reconstitution at 4 °C, from date of receipt.
- 6 months after reconstitution at -20°C to -70°C from date of receipt.

DECLARATION

THIS REAGENT IS FOR IN VITRO LABORATORY TESTING AND RESEARCH USE ONLY. DO NOT USE IT FOR CLINICAL DIAGNOSTICS. DO NOT USE OR INJECT IT IN HUMANS AND ANIMALS.

FOR LABORATORY RESEARCH USE ONLY NOT FOR USE IN HUMANS AND ANIMALS