

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-to-body-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 Z-lines misaligned.^[7]

References

1. Gautel M, et al. (1995). *The EMBO Journal*. **14** (9): 1952–60.
2. Fougousse 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.



Genorise® Recombinant Human MyBPC3 Protein DataSheet

Catalog Number: GR119209

Description

Size: 10 µg

Source: *E coli* derived

Component: Pro2-Ser175

Accession # NP_000247.2.

Predicted Molecular Mass: 19 kDa (monomer)

Specifications

SDS-PAGE: 19 kDa, reducing conditions

Purity: >96%, by SDS-PAGE under reducing conditions and visualized by silver stain.

Formulation: Lyophilized from a 0.2 µm filtered PBS with BSA as a carrier protein.

Preparation and Storage

Reconstitution: Reconstitute at 100 µg/mL in sterile PBS.

Shipping: The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below.

Stability & Storage: Use a manual defrost freezer and avoid repeated freeze thaw cycles.

- 6 months from date of receipt, -20 to -70°C as supplied.
- 3 months, -20 to -70°C under sterile conditions after reconstitution.

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 MICE AND ANIMALS**