

MYBPC3 (h): 293T Lysate: sc-112697

BACKGROUND

MYBPC3 (myosin-binding protein C, cardiac) 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.

REFERENCES

1. Gautel, M., et al. 1995. Phosphorylation switches specific for the cardiac isoform of Myosin binding protein-C: a modulator of cardiac contraction? *EMBO J.* 14: 1952-1960.
2. Bonne, G., et al. 1995. Cardiac Myosin binding protein-C gene splice acceptor site mutation is associated with familial hypertrophic cardiomyopathy. *Nat. Genet.* 11: 438-440.
3. Carrier, L., et al. 1997. Organization and sequence of human cardiac Myosin binding protein C gene (MYBPC3) and identification of mutations predicted to produce truncated proteins in familial hypertrophic cardiomyopathy. *Circ. Res.* 80: 427-434.
4. Yu, B., et al. 1998. Molecular pathology of familial hypertrophic cardiomyopathy caused in the cardiac Myosin binding protein C gene. *J. Med. Genet.* 35: 205-210.
5. Moolman-Smook, J.C., et al. 1998. Identification of a new missense mutation in cardiomyopathy. *J. Med. Genet.* 35: 253-254.
6. Niimura, H., et al. 1998. Mutations in the gene for cardiac Myosin-binding protein C and late-onset familial hypertrophic cardiomyopathy. *N. Engl. J. Med.* 338: 1248-1257.
7. Moolman-Smook, J.C., et al. 1999. The origins of hypertrophic cardiomyopathy-causing mutations in two South African subpopulations: a unique profile of both independent and founder events. *Am. J. Hum. Genet.* 65: 1308-1320.
8. Richard, P., et al. 2003. Hypertrophic cardiomyopathy: distribution of disease genes, spectrum of mutations and implications for a molecular diagnosis strategy. *Circulation* 107: 2227-2232.

CHROMOSOMAL LOCATION

Genetic locus: MYBPC3 (human) mapping to 11p11.2.

PRODUCT

MYBPC3 (h): 293T Lysate represents a lysate of human MYBPC3 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

APPLICATIONS

MYBPC3 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive MYBPC3 antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

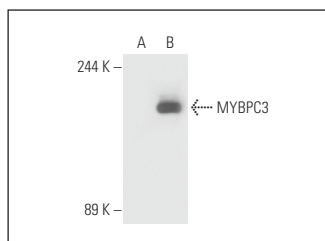
MYBPC3 (G-7): sc-137237 is recommended as a positive control antibody for Western Blot analysis of enhanced human MYBPC3 expression in MYBPC3 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

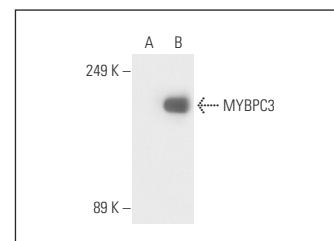
To ensure optimal results, the following support reagents are recommended:

1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

DATA



MYBPC3 (G-7): sc-137237. Western blot analysis of MYBPC3 expression in non-transfected: sc-117752 (A) and human MYBPC3 transfected: sc-112697 (B) 293T whole cell lysates.



MYBPC3 (E-7): sc-137180. Western blot analysis of MYBPC3 expression in non-transfected: sc-117752 (A) and human MYBPC3 transfected: sc-112697 (B) 293T whole cell lysates.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.