

DMPK (9-RY26): sc-134319

BACKGROUND

Myotonic dystrophy protein kinase (DMPK) is a multi-domain protein kinase found in muscle that is activated in response to G protein second messengers and proteolysis. DMPK is implicated in myotonic muscular dystrophy (DM), an autosomal dominant-inherited disorder that predominately affects skeletal and cardiac muscle and causes defects in cardiac conduction. DM arises through expansion of CTG repeats in the 3'-UTR of the DMPK gene. Mutant DMPK transcripts with an extended region of CUG repeats are retained in the nucleus. These transcripts also influence the expression of the DM locus-associated homeodomain protein (DMAHP)/SIX5, to mediate in part the DM phenotype. Other substrates for DMPK include myogenin, L-type calcium channels and phospholemman (PLM).

REFERENCES

- Roberts, R., et al. 1997. Altered phosphorylation and intracellular distribution of a (CUG)_n triplet repeat RNA-binding protein in patients with myotonic dystrophy and in myotonin protein kinase knockout mice. *Proc. Natl. Acad. Sci. USA* 94: 13221-13226.
- Berul, C.I., et al. 1999. DMPK dosage alterations result in atrioventricular conduction abnormalities in a mouse myotonic dystrophy model. *J. Clin. Invest.* 103: R1-R7.
- Mounsey, J.P., et al. 2000. Phospholemman is a substrate for myotonic dystrophy protein kinase. *J. Biol. Chem.* 275: 23362-23367.
- Bush, E.W., et al. 2000. Myotonic dystrophy protein kinase domains mediate localization, oligomerization, novel catalytic activity, and autoinhibition. *Biochemistry* 39: 8480-8490.
- Mankodi, A., et al. 2000. Myotonic dystrophy in transgenic mice expressing an expanded CUG repeat. *Science* 289: 1769-1773.
- Inukai, A., et al. 2000. Reduced expression of DMAHP/SIX5 gene in myotonic dystrophy muscle. *Muscle Nerve* 23: 1421-1426.

CHROMOSOMAL LOCATION

Genetic locus: DMPK (human) mapping to 19q13.32.

SOURCE

DMPK (9-RY26) is a mouse monoclonal antibody raised against an internal region of DMPK of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

PROTOCOLS

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

APPLICATIONS

DMPK (9-RY26) is recommended for detection of DMPK of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for DMPK siRNA (h): sc-38993, DMPK shRNA Plasmid (h): sc-38993-SH and DMPK shRNA (h) Lentiviral Particles: sc-38993-V.

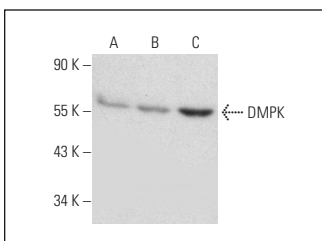
Molecular Weight of DMPK: 45-70 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, K-562 whole cell lysate: sc-2203 or Jurkat whole cell lysate: sc-2204.

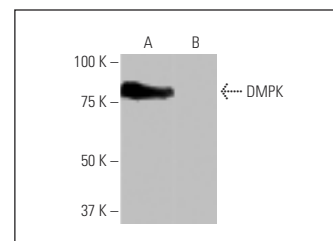
RECOMMENDED SUPPORT REAGENTS

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



DMPK (9-RY26): sc-134319. Western blot analysis of DMPK expression in HeLa (A), K-562 (B) and Jurkat (C) whole cell lysates.



DMPK (9-RY26): sc-134319. Western blot analysis of DMPK expression in human DMPK transfected (A) and non-transfected (B) 293T whole cell lysates.

SELECT PRODUCT CITATIONS

- Provenzano, C., et al. 2017. CRISPR/Cas9-mediated deletion of CTG expansions recovers normal phenotype in myogenic cells derived from myotonic dystrophy 1 patients. *Mol. Ther. Nucleic Acids* 9: 337-348.
- García-Puga, M., et al. 2020. Myotonic dystrophy type 1 cells display impaired metabolism and mitochondrial dysfunction that are reversed by metformin. *Aging* 12: 6260-6275.

RESEARCH USE

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