SANTA CRUZ BIOTECHNOLOGY, INC.

PABPN1 (H-46): sc-67017

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The Power to Question

BACKGROUND

Oculopharyngeal muscular dystrophy (OPMD), an autosomal dominant late-onset progressive disease, generally presents in patients 50-70 years of age with dysphagia, ptosis and proximal limb weakness. OPMD is caused by the abnormal expansion of a (GCG)n trinucleotide repeat in the coding region of the polyadenylate binding protein nuclear 1 (PABPN1, also designated PABP2) gene. In the wildtype form of PABPN1, (GCG)6 codes for the first six alanines in a homopolymeric stretch of ten alanines. In most individuals with OPMD, this (GCG)6 repeat is expanded to (GCG)8-13, leading to a stretch of 12-17 alanines in mutant PABPN1. Mutated PABPN1 forms aggregates consisting of tubular filaments within the nuclei of skeletal muscle fibers. The PABPN1 protein contains two RNA binding domains, a ribonucleoprotein-type RNA binding domain (RNP domain) and an arginine-rich C-terminal domain, which promotes self-association of PABPN1 and cooperative binding to RNA.

REFERENCES

1. Scheuermann, T., et al. 2003. Trinucleotide expansions leading to an extended poly-L-alanine segment in the poly(A)-binding protein PABPN1 cause fibril formation. Protein Sci. 12: 2685-2692.
2. Kuhn, U., et al. 2003. The RNA binding domains of the nuclear poly(A)-binding protein. J. Biol. Chem. 278: 16916-16925.
3. Hino, H., et al. 2004. Myopathy phenotype in transgenic mice expressing mutated PABPN1 as a model of oculopharyngeal muscular dystrophy. Hum. Mol. Genet. 13: 181-190.
4. Davies, J.E., et al. 2005. Doxycycline attenuates and delays toxicity of the oculopharyngeal muscular dystrophy mutation in transgenic mice. Nat. Med. 11: 672-677.
5. Tavanez, J.P., et al. 2005. *In vivo* aggregation properties of the nuclear poly(A)-binding protein PABPN1. RNA 11: 752-762.

CHROMOSOMAL LOCATION

Genetic locus: PABPN1 (human) mapping to 14q11.2; Pabpn1 (mouse) mapping to 14 C3.

SOURCE

PABPN1 (H-46) is a rabbit polyclonal antibody raised against amino acids 261-306 mapping at the C-terminus of PABPN1 of human origin.

PRODUCT

Each vial contains 200 µg IgG 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 or our catalog for detailed protocols and support products.

APPLICATIONS

PABPN1 (H-46) is recommended for detection of PABPN1 of mouse, rat and 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)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

PABPN1 (H-46) is also recommended for detection of PABPN1 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for PABPN1 siRNA (h): sc-44819, PABPN1 siRNA (m): sc-44820, PABPN1 shRNA Plasmid (h): sc-44819-SH, PABPN1 shRNA Plasmid (m): sc-44820-SH, PABPN1 shRNA (h) Lentiviral Particles: sc-44819-V and PABPN1 shRNA (m) Lentiviral Particles: sc-44820-V.

Molecular Weight of PABPN1: 50 kDa.

Positive Controls: THP-1 nuclear extract: sc-24963, RAW 264.7 nuclear extract: sc-24961 or MCF7 nuclear extract: sc-2149.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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