

PABPN1 (K-18): sc-33009

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 (K-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near 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.

Blocking peptide available for competition studies, sc-33009 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

PABPN1 (K-18) 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), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 (K-18) is also recommended for detection of PABPN1 in additional species, including equine, 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.

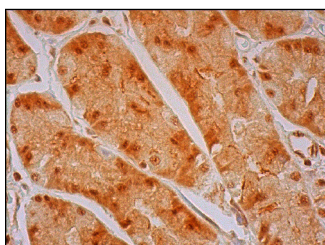
Molecular Weight of PABPN1: 25 kDa.

Positive Controls: MCF7 nuclear extract: sc-2149, THP-1 cell lysate: sc-2238 or NIH/3T3 nuclear extract: sc-2138.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



PABPN1 (K-18): sc-33009. Immunoperoxidase staining of formalin fixed, paraffin-embedded human upper stomach tissue showing nuclear and cytoplasmic staining of glandular cells.

RESEARCH USE

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

PROTOCOLS

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