

PABPN1 (G-17): sc-33007

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.

CHROMOSOMAL LOCATION

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

SOURCE

PABPN1 (G-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-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-33007 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

PABPN1 (G-17) 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), 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 (G-17) is also recommended for detection of PABPN1 in additional species, including 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.

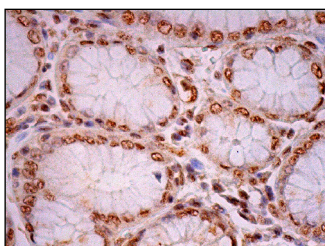
Molecular Weight of PABPN1: 50 kDa.

Positive Controls: NIH/3T3 nuclear extract: sc-2138, RAW 264.7 nuclear extract: sc-24961 or THP-1 nuclear extract: sc-24963.

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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. 4) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



PABPN1 (G-17): sc-33007. Immunoperoxidase staining of formalin fixed, paraffin-embedded human lower stomach tissue showing nuclear staining of glandular cells.

SELECT PRODUCT CITATIONS

- Zhang, K., et al. 2011. RNA interference targeting slug increases cholangiocarcinoma cell sensitivity to cisplatin via upregulating PUMA. *Int. J. Mol. Sci.* 12: 385-400.
- Geibler, V., et al. 2013. The RNA helicase Ddx5/p68 binds to hUpf3 and enhances NMD of Ddx17/p72 and Smg5 mRNA. *Nucleic Acids Res.* 41: 7875-7888.

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.