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SANTA CRUZ BIOTECHNOLOGY, INC.

PABPN1 siRNA (h): sc-44819

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.

PRODUCT

PABPN1 siRNA (h) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see PABPN1 shRNA Plasmid (h): sc-44819-SH and PABPN1 shRNA (h) Lentiviral Particles: sc-44819-V as alternate gene silencing products.

For independent verification of PABPN1 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-44819A, sc-44819B and sc-44819C.

PROTOCOLS

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

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNAses and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

PABPN1 siRNA (h) is recommended for the inhibition of PABPN1 expression in human cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 µM in 66 µl. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor PABPN1 gene expression knockdown using RT-PCR Primer: PABPN1 (h)-PR: sc-44819-PR (20 µl). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

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