# C9orf72 siRNA (h): sc-92761



The Power to Question

### **BACKGROUND**

C9orf72 is a 481 amino acid cytoplasmic and nuclear protein that exists as two alternatively spliced isoforms. Both isoforms of C9orf72 are widely expressed, including expression in kidney, lung, liver, heart, testis, cerebellum and frontal cortex. Defects in C9orf72 are the cause of frontotemporal dementia and/or amyotrophic lateral sclerosis (FTDALS), an autosomal dominant neurodegenerative disorder characterized by adult onset of frontotemporal dementia and/or amyotrophic lateral sclerosis. Frontotemporal dementia is characterized by frontal and temporal lobe atrophy associated with neuronal loss, gliosis and dementia, while amyotrophic lateral sclerosis is characterized by the death of motor neurons in the brain, brainstem and spinal cord, resulting in fatal paralysis.

### **REFERENCES**

- DeJesus-Hernandez, M., et al. 2011. Expanded GGGGCC hexanucleotide repeat in noncoding region of C9orf72 causes chromosome 9p-linked FTD and ALS. Neuron 72: 245-256.
- 2. Renton, A.E., et al. 2011. A hexanucleotide repeat expansion in C9orf72 is the cause of chromosome 9p21-linked ALS-FTD. Neuron 72: 257-268.
- Mahoney, C.J., et al. 2012. Frontotemporal dementia with the C9orf72 hexanucleotide repeat expansion: clinical, neuroanatomical and neuropathological features. Brain 135: 736-750.
- 4. Chiò, A., et al. 2012. Clinical characteristics of patients with familial amyotrophic lateral sclerosis carrying the pathogenic GGGGCC hexanucleotide repeat expansion of C9orf72. Brain 135: 784-793.
- Whitwell, J.L., et al. 2012. Neuroimaging signatures of frontotemporal dementia genetics: C9orf72, Tau, progranulin and sporadics. Brain 135: 794-806.
- Millecamps, S., et al. 2012. Phenotype difference between ALS patients with expanded repeats in C9orf72 and patients with mutations in other ALS-related genes. J. Med. Genet. 49: 258-263.
- 7. Traynor, B.J. 2012. Road to the chromosome 9p-linked ALS/FTD locus. J. Neurol. Neurosurg. Psychiatry 83: 356-357.

# CHROMOSOMAL LOCATION

Genetic locus: C9orf72 (human) mapping to 9p21.2.

### **PRODUCT**

C9orf72 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  $\mu M$  solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see C9orf72 shRNA Plasmid (h): sc-92761-SH and C9orf72 shRNA (h) Lentiviral Particles: sc-92761-V as alternate gene silencing products.

For independent verification of C9orf72 (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-92761A, sc-92761B and sc-92761C.

#### 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  $\mu$ l of the RNAse-free water provided. Resuspension of the siRNA duplex in 330  $\mu$ l of RNAse-free water makes a 10  $\mu$ M solution in a 10  $\mu$ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

### **APPLICATIONS**

C9orf72 siRNA (h) is recommended for the inhibition of C9orf72 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 C9orf72 gene expression knockdown using RT-PCR Primer: C9orf72 (h)-PR: sc-92761-PR (20  $\mu$ l, 563 bp). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

## **SELECT PRODUCT CITATIONS**

1. Leskelä, S., et al. 2018. Interrelationship between the levels of C9orf72 and Amyloid- $\beta$  protein precursor and Amyloid- $\beta$  in human cells and brain samples. J. Alzheimers Dis. 62: 269-278.

## **RESEARCH USE**

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

### **PROTOCOLS**

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

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