

C9orf72 (S-14): sc-138763

BACKGROUND

C9orf72 is a 481 amino acid cytoplasmic and nuclear protein that exists as 2 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

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- Coppo, P., et al. 2006. Bcr-Abl activates Stat3 via JAK and MEK pathways in human cells. *Br. J. Haematol.* 134: 171-179.
- Zheng, X., et al. 2006. Bcr and its mutants, the reciprocal t(9;22)-associated Abl/Bcr fusion proteins, differentially regulate the cytoskeleton and cell motility. *BMC Cancer* 7: 262.
- Burmeister, T., et al. 2007. Atypical Bcr-Abl mRNA transcripts in adult acute lymphoblastic leukemia. *Haematologica* 92: 1699-1702.
- Cottin, V., et al. 2007. Pulmonary vascular manifestations of hereditary hemorrhagic telangiectasia (Rendu-Osler disease). *Respiration* 74: 361-378.
- Fernandez-L, A., et al. 2007. Gene expression fingerprinting for human hereditary hemorrhagic telangiectasia. *Hum. Mol. Genet.* 16: 1515-1533.

CHROMOSOMAL LOCATION

Genetic locus: C9orf72 (human) mapping to 9p21.2; 3110043021Rik (mouse) mapping to 4 A5.

SOURCE

C9orf72 (S-14) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of C9orf72 of human origin.

PRODUCT

Each vial contains 100 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-138763 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.

PROTOCOLS

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

APPLICATIONS

C9orf72 (S-14) is recommended for detection of C9orf72 of human origin, 3110043021Rik of mouse origin and the corresponding rat homolog by Western Blotting (starting dilution 1:100, dilution range 1:50-1:500), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:25, dilution range 1:25-1:250) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with other C9orf family members.

C9orf72 (S-14) is also recommended for detection of C9orf72 in additional species, including equine, canine and avian.

Suitable for use as control antibody for C9orf72 siRNA (h): sc-92761, 3110043021Rik siRNA (m): sc-108907, C9orf72 shRNA Plasmid (h): sc-92761-SH, 3110043021Rik shRNA Plasmid (m): sc-108907-SH, C9orf72 shRNA (h) Lentiviral Particles: sc-92761-V and 3110043021Rik shRNA (m) Lentiviral Particles: sc-108907-V.

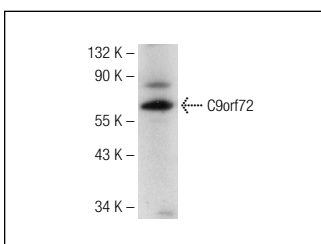
Molecular Weight of C9orf72 isoforms: 54/25 kDa.

Positive Controls: SK-N-SH cell lysate: sc-2410.

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.

DATA



C9orf72 (S-14): sc-138763. Western blot analysis of C9orf72 expression in SK-N-SH whole cell lysate.

SELECT PRODUCT CITATIONS

- 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.

RESEARCH USE

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