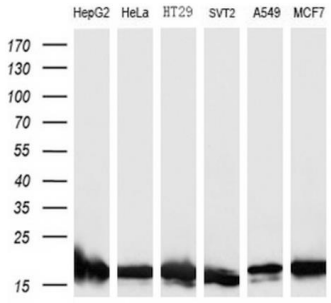
Amyotrophic Lateral Sclerosis

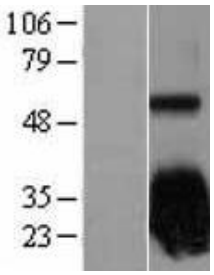
Amyotrophic Lateral Sclerosis (ALS) is described as a neurodegenerative disease and characterized by degeneration of upper and lower motor neurons. ALS is specified by mid-to-late-life onset, selective neuronal death and the formation of protein deposits in affected neuronal tissues similar to other neurodegenerative diseases, like Alzheimer's disease, Parkinson's disease and Huntington's disease. Sporadic cases of ALS occur with no family history of ALS. Familial ALS is due to inherited mutations spread throughout the SOD1 polypeptide.

Superoxide dismutase (SOD) is an antioxidant enzyme and as such part of the defense system against reactive oxygen species. SOD catalyses the dismutation reaction of the superoxide radical anion (O2-) to hydrogen peroxide (H2O2) through glutathione reductase and catalase. Several classes of SODs have been identified, which include the intracellular copper/zinc SOD (**Cu/Zn-SOD, SOD1**), the mitochondrial manganese SOD (**Mn-SOD, SOD2**) and the extracellular copper/zinc SOD (**ECSOD, SOD3**).

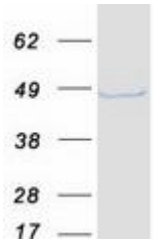
SOD1 is found in all eukaryotic species as a homodimeric 32 kDa enzyme, which contains one Cu- and one Zn-ion each. SOD1 neutralizes supercharged oxygen molecules (superoxide radicals), which can damage cells if their levels are not controlled. The mechanism through which mutant SOD1 causes ALS is not known, but instability, misfolding and aggregation of mutant SOD1 is hypothesized. Biochemical markers of SOD1 misfolding have been noticed as early indicators of ALS. One such marker is the appearance of detergent insoluble precipitates of SOD1 which correlate well with disease onset and progression.



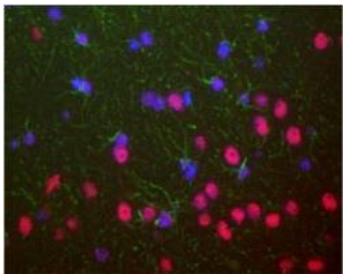
SOD1 antibody, cat# TA500495. Western blot with of extracts (35ug) from 6 different cell lines.



Expression validated SOD2 ORF clone, cat# RC202330, Western validation with an anti-DDK antibody (SKU TA50011-100).
L: Control HEK293 lysate R: Over-expression lysate.



Recombinant TDP43 protein purified from HEK293 cells, cat# TP310639.



TA309896 staining of TDP43 (red) in formalin fixed adult rat hippocampus. Hippocampal neuron nuclei are stained strongly. GFAP (green) shows the processes of astrocytic glial cells. Nuclei of all cells are revealed with DAPI (blue).

The cellular protein **TAR DNA-binding protein 43 (TARDBP, TDP43)** functions as a DNA-binding protein and specifically binds to the TAR DNA sequence motifs of HIV. TARDBP protein has been identified as a major pathological protein of sporadic and familial frontotemporal lobar degeneration with ubiquitin-positive, tau-negative inclusions with or without motor neuron disease. Therefore, TARDBP defines a novel class of neurodegenerative diseases called TARDBP proteinopathies.

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Products for ALS research

Gene/Protein	Description	SKU
SOD1	anti hu SOD1 mouse monocl. antibody; for WB, IHC, FC	TA500495
SOD1	anti hu, ms, rt SOD1 rabbit polycl. antibody; for WB, IHC	TA326813
SOD1	hu SOD1 ORF clone, Myc-DDK tagged	RC200725
SOD1	hu SOD1 lentiviral ORF clone, Myc-DDK tagged	RC200725L2
SOD1	Hu SOD1 lentiviral ORF clone, Myc-DDK tagged, mam. resistance	RC200725L4
SOD1	hu SOD1 lentiviral particles	RC200725L2V
SOD1	hu SOD1 lentiviral particles	RC200725L4V
SOD1	hu SOD1 recomb. protein, mammalian expressed	TP300725
SOD1	hu SOD1 siRNA	SR304518
SOD1	hu SOD1 shRNA in lentiviral vector	TL309191
SOD1	hu SOD1 shRNA as lentiviral particles	TL309191V
SOD1	hu SOD1 CRISPR kit	KN400725
SOD1	hu SOD1 ELISA kit	EA100096
SOD2	anti hu, dg SOD2 mouse monocl. antibody; for WB	TA501887
SOD2	anti hu, ms, rt SOD2 rabbit polycl. antibody; for WB, IHC	TA327034
SOD2	hu SOD2 ORF clone, Myc-DDK tagged	RC202330
SOD2	hu SOD2 lentiviral ORF clone, Myc-DDK tagged	RC202330L2
SOD2	Hu SOD2 lentiviral ORF clone, Myc-DDK tagged, mam. resistance	RC202330L4
SOD2	hu SOD2 lentiviral particles	RC202330L2V
SOD2	hu SOD2 lentiviral particles	RC202330L4V
SOD2	hu SOD2 recomb. protein, mammalian expressed	TP302330
SOD2	hu SOD2 siRNA	SR304519
SOD2	hu SOD2 shRNA in lentiviral vector	TL309190
SOD2	hu SOD2 shRNA as lentiviral particles	TL309190V
SOD2	hu SOD2 CRISPR kit	KN402330
SOD3	anti hu, ms, rt SOD3 mouse monocl. antibody; for IF	TA326420
SOD3	anti hu, ms,rt SOD3 rabbit polycl. antibody; for WB	TA326421
SOD3	hu SOD3 ORF clone, Myc-DDK tagged	RC204156
SOD3	hu SOD3 lentiviral ORF clone, Myc-DDK tagged	RC204156L2
SOD3	Hu SOD3 lentiviral ORF clone, Myc-DDK tagged, mam. resistance	RC204156L4
SOD3	hu SOD3 lentiviral particles	RC204156L2V
SOD3	hu SOD3 lentiviral particles, incl. mam. resistance	RC204156L4V
SOD3	hu SOD3 recomb. protein, mammalian expressed	TP304156
SOD3	hu SOD3 siRNA	SR304520
SOD3	hu SOD3 shRNA in lentiviral vector	TL316735
SOD3	hu SOD3 shRNA as lentiviral particles	TL316735V
SOD3	hu SOD3 CRISPR kit	KN404156
TDP43	anti hu, ms, rt TDP43 mouse monocl. antibody; for IF	TA309896
TDP43	anti hu, ms TDP43 rabbit polycl. antibody; for WB, IHC, IF	TA324524
TDP43	hu SOD3 ORF clone, Myc-DDK tagged	RC210639
TDP43	hu SOD3 lentiviral ORF clone, Myc-DDK tagged	RC210639L2
TDP43	Hu SOD3 lentiviral ORF clone, Myc-DDK tagged, mam. resistance	RC210639L4
TDP43	hu SOD3 lentiviral particles	RC210639L2V
TDP43	hu SOD3 lentiviral particles, incl. mam. resistance	RC210639L4V
TDP43	hu TDP43 recomb. protein, mammalian expressed	TP310639
TDP43	hu TDP43 siRNA	SR308276
TDP43	hu SOD3 shRNA in lentiviral vector	TL308946
TDP43	hu SOD3 shRNA as lentiviral particles	TL308946V
TDP43	hu TDP43 CRISPR kit	KN410639