

## Product datasheet

# MISFOLDED SOD1 MOUSE MONOCLONAL ANTIBODY (C4F6)

SKU: MM-0070-2-P

200 µL

#### **OVERVIEW**

## Clonality:

Monoclonal

#### Host:

Mouse

## Reactivity:

Human, Mouse (IF)

## Application:

IF, IHC, IP, WB

#### Target:

Misfolded SOD1

## Target background:

Superoxide dismutase 1 (SOD1) is a soluble cytoplasmic and mitochondrial intermembrane space protein. SOD1 binds copper and zinc ions and is one of three isozymes responsible for destroying free superoxide radicals in the body.

Mutations in SOD1 cause familial amyotrophic lateral sclerosis type 1 (ALS1).

These mutations have been linked to accumulation of harmful superoxide radicals, promotion of apoptosis, formation of aggregates of misfolded superoxide dismutase which are toxic and the continued stimulation of nerve cells that causes them to burn out and die.

## Target alias:

Superoxide dismutase [Cu-Zn], Superoxide dismutase 1, hSOD1, SOD1

## Immunogen:

Recombinant human SOD1 G93A APO

## Specificity:

The antibody recognizes misfolded forms of mutant human SOD1 protein. IF: WT, G93A, G85R, G127X, G37R and A4V; WB (denatured): WT, G93A, D90A, G37R and A4V; WB (native): G93A; IHC: WT, G93A and G37R.

## Clone ID:

C4F6

## Isotype:

lgG2a

## Preservative:

Contains 1% BSA and 0.02% sodium azide.

## Format:

Lyophilized protein G purified in PBS pH7.4

## Recommend starting dilution:

If reconstituted with deionized water in 200  $\mu$ L: IF 1:50 to 1:500; WB 1:250; IP 6  $\mu$ L / 40  $\mu$ L (Protein G beads / 300  $\mu$ g of protein lysate). Optimal dilution has to be determined by the user.

## Limitations:

Research Use Only

## References:

- 1.-Da Cruz S Misfolded SOD1 is not a primary component of sporadic ALS.
- 2.-Pickles S ALS-linked misfolded SOD1 species have divergent impacts on mitochondria.
- 3.-Ayers JI Conformational specificity of the C4F6 SOD1 antibody; low frequency of reactivity in sporadic ALS cases.
- 4.-Gros-Louis F Intracerebroventricular infusion of monoclonal antibody or its derived Fab fragment against misfolded forms...
- 5.-Pickles S Mitochondrial damage revealed by immunoselection for ALS-linked misfolded SOD1.
- 6.-Roberts BR Oral treatment with Cu(II)(atsm) increases mutant SOD1 in vivo but protects motor neurons and improves the...
- 7.-Brown HH and Borchelt DR Analysis of mutant SOD1 electrophoretic mobility by Blue Native gel electrophoresis; evidence...
- 8.-Bosco DA Wild-type and mutant SOD1 share an aberrant conformation and a common pathogenic pathway in ALS.
- 9.-Chhangani D Mahogunin ring finger 1 confers cytoprotection against mutant SOD1 aggresomes and is defective in an ALS...
- 10.-Jouroukhin Y NAP (davunetide) modifies disease progression in a mouse model of severe neurodegeneration: protection...
- 11.-Nizzardo M Morpholino-mediated SOD1 reduction ameliorates an amyotrophic lateral sclerosis disease phenotype.
- 12.-Patel P Adeno-associated virus-mediated delivery of a recombinant singlechain antibody against misfolded superoxide...
- 13.-Redler RL Non-native soluble oligomers of Cu/Zn superoxide dismutase (SOD1) contain a conformational epitope linked...
- 14.-Sábado J Adverse effects of a SOD1-peptide immunotherapy on SOD1 G93A mouse slow model of amyotrophic lateral sclerosis.
- 15.-Xu G Substantially elevating the levels of αB-crystallin in spinal motor neurons of mutant SOD1 mice does not...
- 16.-Xu G Direct and indirect mechanisms for wild-type SOD1 to enhance the toxicity of mutant SOD1 in bigenic transgenic mice.
- 17.-Okamoto Y Colocalization of 14-3-3 proteins with SOD1 in Lewy body-like hyaline inclusions in familial amyotrophic...
- 18.-Urushitani M Therapeutic effects of immunization with mutant superoxide dismutase in mice models of amyotrophic...

## Storage:

Lyophilized antibodies can be kept at 4°C for up to 3 months and should be kept at -20°C for long-term storage (2 years). To avoid freeze-thaw cycles, reconstituted antibodies should be aliquoted before freezing for long-term (1 year) storage (-80°C) or kept at 4°C for short-term usage (2 months). For maximum recovery of product, centrifuge the original vial prior to removing the cap. Further dilutions can be made with the assay buffer. After the maximum long-term storage period (2 years lyophilized or 1 year reconstituted) antibodies should be tested in your assay with a standard sample to verify if you have noticed any decrease in their efficacy.

## Image:

C4F6 G93A Sc P75 (5x) G93A Sc P75 (10x) G93A Sc P75 (20x) Tg-hWTov Sc P75 (5x)

The spinal cord sections of transgenic mice SOD1G93A show the detection of misfolded SOD1 in the motor neurons. This immunodetection does not appear in the transgenic mice that reproduce normal human SOD1 (Tg-hWTov Sc). MM-0070-2-P: 1:1000

