

## Product datasheet

### MISFOLDED SOD1 MOUSE MONOCLONAL ANTIBODY (A5C3)

**SKU:** MM-0070-3-P

200 µL

#### OVERVIEW

**Clonality:**

Monoclonal

**Host:**

Mouse

**Reactivity:**

Human

**Application:**

WB, IP, IF

**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: G93A, G85R, and G37R; WB (denatured): G93A, G127X and G37R; WB (native): G37R; IP: G93A, G127X and G37R.

**Clone ID:**

A5C3

**Isotype:**

IgG1

**Preservative:**

None

**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 5  $\mu$ L / 40  $\mu$ L (Protein G beads / 300  $\mu$ g of protein lysate). Optimal dilution must be determined by the user.

**Limitations:**

Research Use Only

### References:

- 1.-Pickles S - ALS-linked misfolded SOD1 species have divergent impacts on mitochondria.
- 2.-Gros-Louis F - Intracerebroventricular infusion of monoclonal antibody or its derived Fab fragment against misfolded forms of SOD1 mutant delays mortality in a mo...
- 3.-Vande Velde C - Misfolded SOD1 associated with motor neuron mitochondria alters mitochondrial shape and distribution prior to clinical onset.
- 4.-Audet JN - Methylene blue administration fails to confer neuroprotection in two amyotrophic lateral sclerosis mouse models.
- 5.-Lee JC - Replacement of microglial cells using Clodronate liposome and bone marrow transplantation in the central nervous system of SOD1(G93A) transgenic mi...
- 6.-Parone PA - Enhancing mitochondrial calcium buffering capacity reduces aggregation of misfolded SOD1 and motor neuron cell death without extending survival in ...
- 7.-Roberts BR - Oral treatment with Cu(II)(atsm) increases mutant SOD1 in vivo but protects motor neurons and improves the phenotype of a transgenic mouse model of...

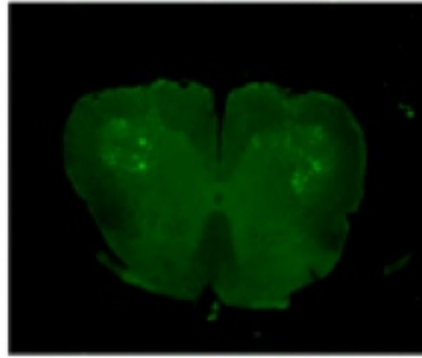
### Storage:

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.

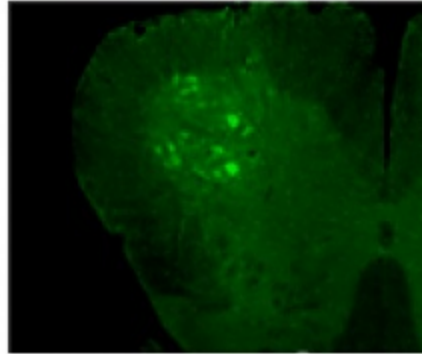
Image:

## A5C3

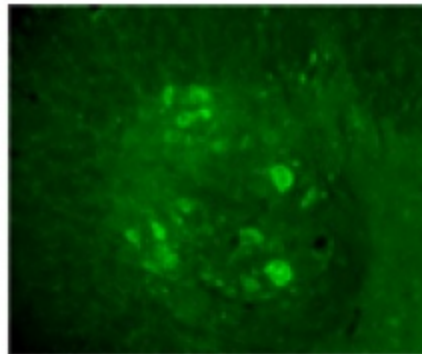
G93A Sc  
P75 (5x)



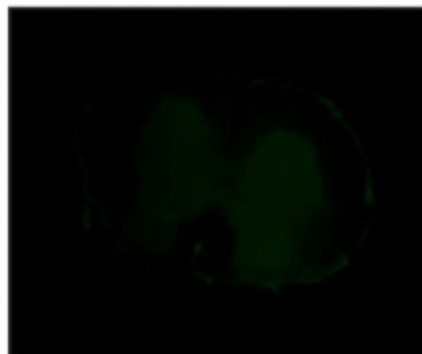
G93A Sc  
P75 (10x)



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-0070-3-P: 1:1000

