abcam

Product datasheet

Recombinant human Superoxide Dismutase 1 protein ab201408

Description

Product name	Recombinant human Superoxide Dismutase 1 protein
Biological activity	Fully biologically active when compared to standard. The potency per mg was tested by Pyrogallic Acid method and was found to be more than 10,000Units/mg.
Purity	> 95 % SDS-PAGE. >95% by HPLC analysis.
Endotoxin level	< 1.000 Eu/µg
Expression system	Escherichia coli
Accession	P00441
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MATKAVCVLKGDGPVQGIINFEQKESNGPVKVWGSIKGLT EGLHGFHVHE FGDNTAGCTSAGPHFNPLSRKHGGPKDEERHVGDLGNV TADKDGVADVSI EDSVISLSGDHCIIGRTLVVHEKADDLGKGGNEESTKTGN AGSRLACGVI GIAQ
Predicted molecular weight	31 kDa
Amino acids	1 to 154
Additional sequence information	Homodimer, non-glycosylated polypeptide chain containing 2 x 154 amino acids.

Specifications

Our **Abpromise guarantee** covers the use of **ab201408** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Functional Studies HPLC SDS-PAGE

Form	Lyophilized

Preparation and Storage

Stability and Storage	Shipped at 4°C. Store at -20°C long term. Avoid freeze / thaw cycle. pH: 7.40 Constituent: 100% PBS Lyophilised from a 0.2μm filtered solution. This product is an active protein and may elicit a biological response in vivo, handle with caution.
Reconstitution	Briefly centrifuge the vial prior to opening to bring the contents to the bottom. Reconstitute in sterile distilled water or aqueous buffer containing 0.1% BSA to a concentration of 0.1-1.0 mg/ml. Stock solutions should be apportioned into working aliquots and stored at -20°C to -70°C. Further dilutions should be made in appropriate buffered solutions.

General Info

Function	Destroys radicals which are normally produced within the cells and which are toxic to biological systems.
Involvement in disease	Defects in SOD1 are the cause of amyotrophic lateral sclerosis type 1 (ALS1) [MIM:105400]. ALS1 is a familial form of amyotrophic lateral sclerosis, a neurodegenerative disorder affecting upper and lower motor neurons and resulting in fatal paralysis. Sensory abnormalities are absent. Death usually occurs within 2 to 5 years. The etiology of amyotrophic lateral sclerosis is likely to be multifactorial, involving both genetic and environmental factors. The disease is inherited in 5-10% of cases leading to familial forms.
Sequence similarities	Belongs to the Cu-Zn superoxide dismutase family.
Post-translational modifications	Unlike wild-type protein, the pathogenic variants ALS1 Arg-38, Arg-47, Arg-86 and Ala-94 are polyubiquitinated by RNF19A leading to their proteasomal degradation. The pathogenic variants ALS1 Arg-86 and Ala-94 are ubiquitinated by MARCH5 leading to their proteasomal degradation. The ditryptophan cross-link at Trp-33 is reponsible for the non-disulfide-linked homodimerization. Such modification might only occur in extreme conditions and additional experimental evidence is required.
Cellular localization	Cytoplasm. The pathogenic variants ALS1 Arg-86 and Ala-94 gradually aggregates and accumulates in mitochondria.

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