

Product datasheet

Recombinant human Superoxide Dismutase 1 protein (Active) ab112193

[4 References](#) [1 Image](#)

Description

Product name	Recombinant human Superoxide Dismutase 1 protein (Active)
Biological activity	This protein is fully biologically active when compared to standard. Activity tests were carried using ab65354 . The activity assay kit showed that the calculated activity was ~40,000 U/mg.
Purity	> 95 % SDS-PAGE. > 95% SDS-PAGE. The Cu/Zn SOD is purified by proprietary chromatographic techniques.
Expression system	Escherichia coli
Accession	<u>P00441</u>
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Predicted molecular weight	17 kDa

Specifications

Our [Abpromise guarantee](#) covers the use of **ab112193** in the following tested applications.

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

Applications	SDS-PAGE Functional Studies
Form	Lyophilized
Additional notes	This product is manufactured by BioVision, an Abcam company and was previously called 4802 Superoxide Dismutase (SOD), human recombinant. 4802-100 is the same size as the 100 µg size of ab112193. Endotoxin Levels: <0.1 ng/mg.

Preparation and Storage

Stability and Storage	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
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pH: 7.40

Constituents: 10.269% Trehalose, 0.727% Dibasic monohydrogen potassium phosphate, 0.248% Monobasic dihydrogen potassium phosphate

This product is an active protein and may elicit a biological response in vivo, handle with caution.

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

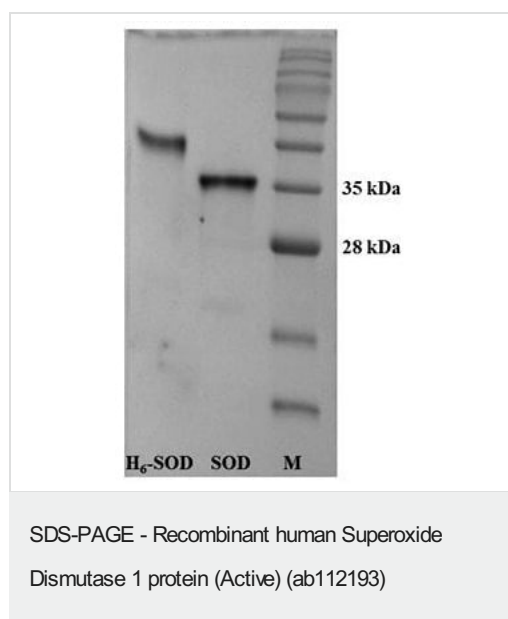
Unlike wild-type protein, the pathogenic variants ALS1 Arg-38, Arg-47, Arg-86 and Ala-94 are polyubiquitinated by RNF19A leading to their proteasomal degradation. The pathogenic variants ALS1 Arg-86 and Ala-94 are ubiquitinated by MARCH5 leading to their proteasomal degradation.

The ditryptophan cross-link at Trp-33 is responsible 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.

Images



20 ug of non-reduced ab112193 on SDS-PAGE, stained with Coomassie Blue after protein migration.

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