

Product datasheet

Recombinant Human Superoxide Dismutase 1 protein ab82649

2 图像

概述

产品名称	Recombinant人Superoxide Dismutase 1 protein
蛋白长度	Full length protein

描述

性质	Recombinant
来源	Escherichia coli
氨基酸序列	
种属	Human

技术指标

Our [Abpromise guarantee](#) covers the use of **ab82649** in the following tested applications.

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

应用	SDS-PAGE Western blot
纯度	> 95 % SDS-PAGE.
形式	Lyophilised

制备和贮存

稳定性和存储	Shipped at 4°C. Upon delivery aliquot. Store at -80°C. Avoid freeze / thaw cycle. Preservative: None Constituents: 20mM HEPES, pH 7.4
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常规信息

功能	Destroys radicals which are normally produced within the cells and which are toxic to biological systems.
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## 疾病相关

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.

## 序列相似性

Belongs to the Cu-Zn superoxide dismutase family.

## 翻译后修饰

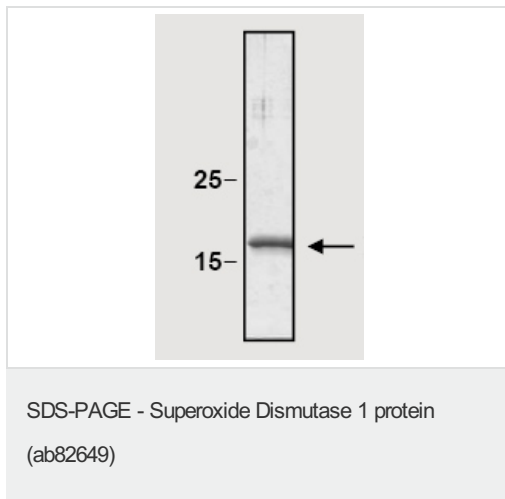
Unlike wild-type protein, the pathogenic variants ALS1 Arg-38, Arg-47, Arg-86 and Ala-94 are polyubiquitinated by RNF19A leading to their proteasomal degradation. The pathogenic variants ALS1 Arg-86 and Ala-94 are ubiquitinated by MARCH5 leading to their proteasomal degradation.

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.

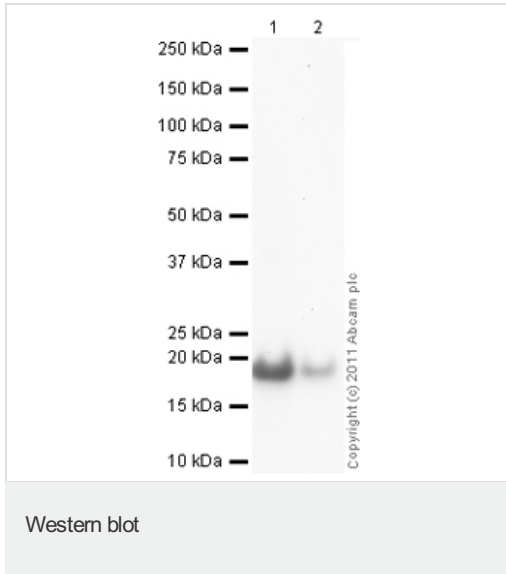
## 细胞定位

Cytoplasm. The pathogenic variants ALS1 Arg-86 and Ala-94 gradually aggregates and accumulates in mitochondria.

## 图片



ab82649 (1 $\mu$ g) on SDS-PAGE.



**All lanes :** Anti-Superoxide Dismutase 1 antibody ([ab88404](#)) at 1 µg/ml

**Lane 1 :** Recombinant Human Superoxide Dismutase 1 protein ([ab82649](#)) at 0.01 µg

**Lane 2 :** Recombinant Human Superoxide Dismutase 1 protein ([ab82649](#)) at 0.001 µg

**Secondary**

Goat Anti-Rabbit IgG H&L (HRP) preadsorbed ([ab97080](#)) at 1/5000 dilution

Developed using the ECL technique

Performed under reducing conditions.

**Exposure time :** 30 seconds

**Please note:** All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

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