abcam

Product datasheet

Recombinant Human Superoxide Dismutase 1 protein ab84642

1 图像

描述

产品名称	重组人Superoxide Dismutase 1蛋白
纯度	> 90 % Densitometry.
表达系统	Escherichia coli
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human

技术指标

Our **Abpromise guarantee** covers the use of **ab84642** in the following tested applications.

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

应用	Western blot SDS-PAGE
形式	Liquid

制备和贮存

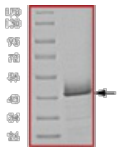
稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 7.50 Constituents: 0.00174% PMSF, 0.00385% DTT, 0.79% Tris HCl, 25% Glycerol (glycerin, glycerine), 0.87% Sodium chloride

常规信息

功能	Destroys radicals which are normally produced within the cells and which are toxic to biological systems.
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 reponsible 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



SDS-PAGE - Recombinant Human Superoxide Dismutase 1 protein (ab84642)

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