abcam

Product datasheet

Recombinant Human Superoxide Dismutase 1 protein ab84642

画像数1

製品の詳細

製品名 Recombinant Human Superoxide Dismutase 1 protein

精製度 > 90 % Densitometry.

発現系 Escherichia coli

タンパク質長 Full length protein

Animal free No

由来 Recombinant

生物種 Human

特性

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.

アプリケーション Western blot

SDS-PAGE

製品の状態 Liquid

前処理および保存

保存方法および安定性 Shipped on dry ice. Upon delivery aliquot and store at -80°C. 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

関連情報

機能 Destroys radicals which are normally produced within the cells and which are toxic to biological

systems.

関連疾患 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.

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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.

配列類似性 Belongs to the Cu-Zn superoxide dismutase family.

翻訳後修飾 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.

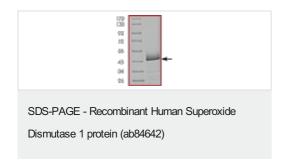
The ditryptophan cross-link at Trp-33 is reponsible for the non-disulfide-linked homodimerization. Such modification might only occur in extreme conditions and additional experimental evidence is

required.

細胞内局在 Cytoplasm. The pathogenic variants ALS1 Arg-86 and Ala-94 gradually aggregates and

accumulates in mitochondria.

画像



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