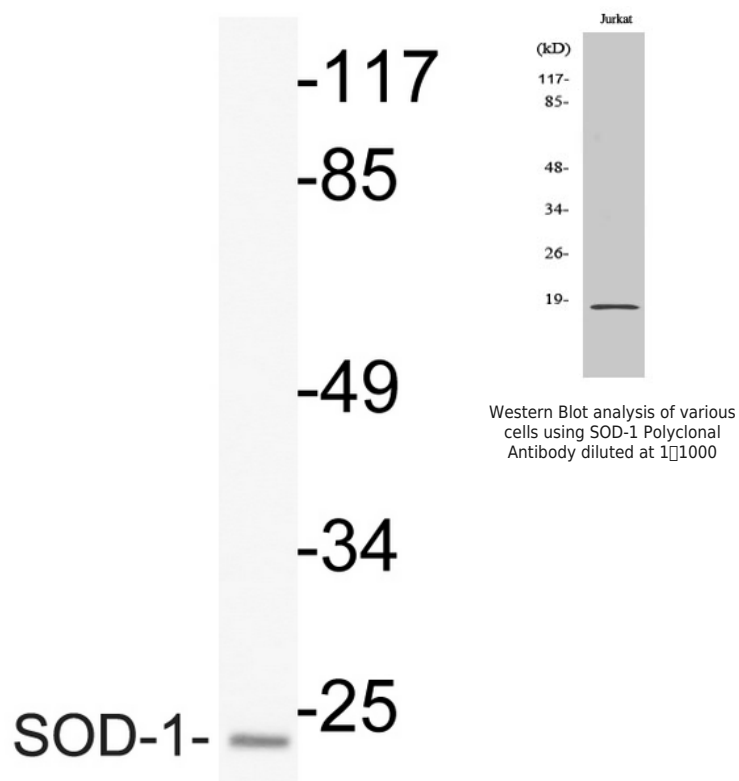


Product name:	SOD-1 Rabbit Polyclonal Antibody
Cat number:	ABN18097
Conjugate:	Unconjugated
Size:	100 ug
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human SOD-1. AA range:36-85
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,ELISA 1:5000-1:20000
Molecular Weight:	~15kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%protective protein.
Storage:	Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.

Background:

The protein encoded by this gene binds copper and zinc ions and is one of two isozymes responsible for destroying free superoxide radicals in the body. The encoded isozyme is a soluble cytoplasmic protein, acting as a homodimer to convert naturally-occurring but harmful superoxide radicals to molecular oxygen and hydrogen peroxide. The other isozyme is a mitochondrial protein. Mutations in this gene have been implicated as causes of familial amyotrophic lateral sclerosis. Rare transcript variants have been reported for this gene. [provided by RefSeq, Jul 2008], catalytic activity: $2 \text{ superoxide} + 2 \text{ H}^{+} = \text{O}_2 + \text{H}_2\text{O}_2$, cofactor: Binds 1 copper ion per subunit, cofactor: Binds 1 zinc ion per subunit, 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, function: Destroys radicals which are normally produced within the cells and which are toxic to biological systems, miscellaneous: The protein (both wild-type and ALS1 variants) has a tendency to form fibrillar aggregates in the absence of the intramolecular disulfide bond or of bound zinc ions. These aggregates may have cytotoxic effects. Zinc binding promotes dimerization and stabilizes the native form, online information: ALS genetic mutations db, online information: Superoxide dismutase entry, PTM: Unlike wild-type protein, the pathogenic variants ALS1 Arg-38, Arg-47, Arg-86 and Ala-94 are polyubiquitinated by RNF19A; which leads to their proteasomal degradation, similarity: Belongs to the Cu-Zn superoxide dismutase family, subunit: Homodimer. The pathogenic variants ALS1 Arg-38, Arg-47, Arg-86 and Ala-94 interact with RNF19A, whereas wild-type protein does not.



Western blot analysis of lysate from Jurkat cells, using SOD-1 antibody.