

Product Information

Anti-Dorfin (N-terminal)

produced in rabbit, affinity isolated antibody

Catalog Number **D0319**

Product Description

Anti-Dorfin (N-terminal) is produced in rabbit using as immunogen a synthetic peptide corresponding to amino acids 1-19 of human Dorfin (GeneID 25897) conjugated to KLH. This sequence is identical in mouse Dorfin and highly conserved (two amino acid substitutions) in rat Dorfin. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-Dorfin (N-terminal) specifically recognizes human and mouse Dorfin by immunoblotting (~105 kDa). Detection of the Dorfin band is specifically inhibited by the Dorfin immunizing peptide.

The ubiquitin-proteasome system (UPS) is involved in the pathogenic mechanisms of several common neurodegenerative diseases including Parkinson's disease (PD), amyotrophic lateral sclerosis (ALS), polyglutamine diseases such as Huntington's disease (HD), and Alzheimer's disease (AD).^{1,2} Misfolded protein accumulation, a probable cause of neurodegenerative diseases, can cause deterioration of various cellular functions leading to neuronal cell death. Dorfin (Double ring-finger protein, RING finger protein 19, RNF19) is an E3 ubiquitin ligase that contains a RING finger/IBR (in between-RING-finger) domain at its N-terminus and mediates E3 ubiquitin ligase activity.³ Dorfin interacts with human E2s UbcH7 and UbcH8. The C-terminus of Dorfin can recognize mutant SOD1 proteins, which cause familial ALS (FALS). Dorfin binds and ubiquitinylates various SOD1 mutants derived from FALS and enhances their degradation, but has no effect on the stability of wild-type SOD1.⁴ Over-expression of Dorfin protects neuronal cells against the toxic effect of mutant SOD1 and reduces SOD1 inclusions. In cultured cells, Dorfin resides in the perinuclear region and forms aggresome-like structures, that contain misfolded ubiquitylated proteins.³ Dorfin has been localized to aggresomes and ubiquitin positive inclusions in several neurodegenerative diseases including: PD, dementia with Lewy bodies, multiple system atrophy and ALS.^{5,6}

Reagent

Supplied as a solution in 0.01 M PBS, pH 7.4, containing 15 mM sodium azide as a preservative.

Antibody concentration: ~1.5 mg/mL

Precautions and Disclaimer

This product is for R&D use only, not for drug, household, or other uses. Please consult the Material Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability

For continuous use, store at 2-8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in "frost-free" freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilutions should be discarded if not used within 12 hours.

Product Profile

Immunoblotting: a working concentration of 1-2 µg/mL is recommended using HEK-293T cells expressing human Dorfin.

Note: In order to obtain the best results using various techniques and preparations, we recommend determining the optimal working dilutions by titration.

References

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3. Niwa, J., et al., *Biochem. Biophys. Res. Commun.*, **281**, 706-713 (2001).
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5. Hishikawa, N., et al., *Am. J. Pathol.*, **163**, 609-619 (2003).
6. Ito, T., et al., *J. Biol. Chem.*, **278**, 29106-29114 (2003).

ER,DXP,PHC 03/08-1