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Product Information

Anti-ATP13A2 (C-terminal region)
produced in rabbit, affinity isolated antibody

Product Number **A3361**

Product Description

Anti-ATP13A2 (C-terminal region) is produced in rabbit using as the immunogen a synthetic peptide corresponding to a sequence at the C-terminal of human ATP13A2 (Gene ID: 23400) conjugated to KLH. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-ATP13A2 (C-terminal region) specifically recognizes human and mouse ATP13A2. The antibody may be used in various immunochemical techniques including immunoblotting (~129 kDa). Staining of the ATP13A2 band by immunoblotting is specifically inhibited by the ATP13A2 immunizing peptide.

Parkinson's disease (PD) is the most common motor neurodegenerative disease, characterized by the progressive loss of dopaminergic neurons from the substantia nigra and the presence of intracellular Lewy bodies. Mutations in several genes have been linked to PD in recent years. This has led to the discovery of pathophysiological pathways including enhanced oxidative stress, protein misfolding and aggregation, and dysfunction of the ubiquitin-proteasome system.[1-3] Mutations in the gene encoding ATP13A2 (ATPase type 13A2, also known as PARK9) a neuronal P-type ATPase of the P_5 subfamily, have been recently identified in an hereditary form of PD associated with dementia, known as Kufor-Rakeb syndrome (KRS).[4-7] KRS is a rare form of hereditary PD with juvenile onset. In addition to typical signs of PD, affected individuals show symptoms of more widespread pyramidal neurodegeneration, including dementia. Wild-type ATP13A2 is located in the lysosome of transiently transfected cells, whereas the unstable truncated mutants are retained in the endoplasmic reticulum and degraded by the proteasome. ATP13A2 shows elevated expression levels in the brains of sporadic PD patients, suggesting a potential role in the more common forms of Parkinson's disease.

Reagent

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

Antibody concentration: ~1.5 mg/mL

Precautions and Disclaimer

For R&D use only. Not for drug, household, or other uses. Please consult the Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability

Store at –20 °C. For continuous use, the product may be stored at 2–8 °C for up to one month. For extended storage, freeze in working aliquots at –20 °C. 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 antibody concentration of 1.5-3.0 μg/mL is recommended using a mouse brain extract (S1 fraction) or HEK-293T cells expressing human ATP13A2.

Note: In order to obtain best results in various techniques and preparations, it is recommended to determine optimal working dilutions by titration.

References

1. Ciechanover, A., *Nature Rev. Mol. Cell Biol.,* **6**, 79-87 (2005).
2. Cookson, M.R., *Ann. Rev. Biochem.,* **74**, 29-52 (2005).
3. Dawson, T.M., and Dawson, V.L., *Science,* **302**, 819-822 (2003).
4. Ramirez, A. et al., *Nature Genet.,* **38**, 1184-1191 (2006).
5. Williams, D.R. et al., *Mov. Disord.,* **20**, 1264-1271 (2005).
6. Wei, J. et al., *J. Biol. Chem.,* **282**, 28904-28914 (2007).
7. Ning, Y.P. et al., *Neurol.,* **70**, 1491-1493 (2008).

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