

## Product Information

### Anti-Ubiquilin-1

produced in rabbit, affinity isolated antibody

Product Number **U7258**

#### Product Description

Anti-Ubiquilin-1 is produced in rabbit using as immunogen a synthetic peptide corresponding to a fragment of human ubiquilin-1 (GeneID 29979) conjugated to KLH. This sequence is not found in rat and mouse ubiquilin-1. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-Ubiquilin-1 specifically recognizes human ubiquilin-1 by immunoblotting (~62 kDa). Staining of the ubiquilin-1 band is specifically inhibited by the immunizing peptide.

The ubiquitin-proteasome system (UPS) is involved in the pathogenic mechanisms of several common neurodegenerative diseases.<sup>1</sup> The accumulation of misfolded proteins, induced by oxidative stress or neurotoxin exposure, leads to protein aggregates and inclusion bodies and a consequent loss of cell function and neuronal cell death. Ubiquilin-1 (also known as UBQLN1, Protein-linking IAP to cytoskeleton 1, PLIC-1, DA41, DSK2, XDRP1) is an ubiquitin-like (UBL) protein that has been shown to play a central role in regulating the proteasomal degradation of various proteins including the presenilins PS1 and PS2.<sup>2-4</sup> UBQLN1 contains UBL and ubiquitin-associated (UBA) domains in its N- and C-termini. Several studies indicate that UBQLN1 may play a general role in neurodegenerative diseases. Genetic variants of UBQLN1 have been associated with increased risk of Alzheimer's disease (AD).<sup>5</sup> Down-regulation of UBQLN1 has been reported to modulate PS1 endoproteolysis along with protein levels of nicastrin and PEN-2 and to increase the rate of APP maturation and trafficking through the secretory pathway leading to increased secretion of sAPP and A $\beta$ .<sup>6</sup> UBQLN1 has been associated with neurofibrillary tangles in AD brain and Lewy bodies in Parkinson's disease (PD).<sup>7</sup> UBQLN1 has been identified as part of polyQ aggregates and in neuronal intranuclear inclusions in a mouse model of Huntington's disease.<sup>8</sup>

#### Reagent

Supplied as a solution in 0.01 M PBS, pH 7.4, containing 1% BSA and 15 mM sodium azide as a preservative.

Antibody concentration: ~0.5 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

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 0.5–1  $\mu$ g/mL is recommended using lysates of HEK-293T cells and COS7 cells expressing human ubiquilin-1.

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

#### References

1. Glickman, M.H., and Ciechanover, A., *Physiol. Rev.*, **82**, 373-428 (2002).
2. Kleijnen, M.F. et al., *Mol. Cell*, **6**, 409-419 (2000).
3. Massey, L.K. et al., *Biochem. J.*, **391**, 513-525 (2005).
4. Bedford, F.K. et al., *Nature Neurosci.*, **4**, 908-916 (2001).

5. Bertram., L. et al., *N. Engl. J. Med.*, **352**, 884-894 (2005).
6. Hiltunen., M. et al., *J. Biol. Chem.*, **281**, 32240-32253 (2006).

7. Mah., A.L. et al., *J. Cell Biol.*, **151**, 847-862 (2000).
8. Doi., H., et al., *FEBS Lett.*, **571**, 171-176 (2004).

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