Mouse monoclonal antibody to Ubiquilin 2

Catalogue No.:	M-1656-100
Description:	Ubiquilin 2 (also known as PLIC2 and Chap1) is a member of the ubiquilin protein family, which regulate the degradation of cellular proteins through proteasome or autophage-like pathways (1, 2, 3). Humans have four ubiquilin genes, each encoding a separate protein referred to as Ubiquilin 1, 2, 3 and 4. All ubiquilins contain an N-terminal ubiquitin-like (UBL) domain and a C-terminal ubiquitin-associated (UBA) domain, while the central part of the molecules are highly variable. The UBL domains bind subunits of the proteasome, and the UBA domains binds to polyubiquitin chains that are typically conjugated onto proteins marked for proteosomal degradation (1). Ubiquilin 2 has a unique region close to the C terminus containing 12 PXX tandem collagen like repeats, where P is proline and X is most cases valine, glycine, isoleucine or threonine. Teepu Siddique and his collaborators have identified mutations in the ubiquilin 2 gene leading to protein point mutations which were important contributors to several forms of amyotrophic lateral sclerosis (ALS) and Frontotemporal lobar degeneration (FTLD). Interestingly, these mutations involved alterations in proline residues in the PXX repeat region (P497H, P497S, P506T, P509S and P525S, ref. 4). Recently, the Lee and Trojanowski group investigated C9orf72 hexanucleotide expansion and ubiquilin 2 pathology in patients with ALS and FTLD by genetic analysis and immunohistochemistry and found distinct ubiquilin 2 pathology in ALS and FTLD-TDP with C9orf72 expansion (5). C9orf72 hexonucleotide expansion is the most commmon cause to date of familial ALS and FTLD (6, 7). Ubiquilin 2 protein is of different molecular size in mouse and human, 638 and 624 amino acids respectively. As a result the mouse protein, endogenously expressed in rodent 3T3 cells, runs on SDS-PAGE and western blots slightly slower than the human protein.
Unit size:	100 ug
Antigen:	Recombinant human ubiquilin 2 expressed and purified from E. coli.
Isotype:	IgG1
Produced in:	Mouse
Applications:	Western Blotting (WB) and Immunocytochemistry (IC). A dilution of 1:1,000 - 1:2,000 is recommended for WB. A dilution of 1:500-1:1,000 is recommended for IC. Biosensis recommends optimal dilutions/concentrations should be determined by the end user.
Specificity:	In primary mouse neuron and glia cell culture, endogenous ubiquilin 2 appears as a weak band at 68kDa in all tranduced and non-transduced cells, indicating low endogenous expression of mouse ubiquilin 2. Strong bands are seen in cells transduced with human wild type or mutant ubiquilin 2. Small proteins which run at 50 kDa in these cells are the fragments of ubiquilin 2. Note, ubiquilin 2 runs at ~66 kDa in human Hela cells and 68 kDa in rodent 3T3 cells. The antibody has also been used successfully for immunocytochemistry.
Species Against:	Human and mouse
Antibody Against:	Ubiquilin 2 (PLIC2; Chap1)
Form:	Lyophilized from PBS containing 5% trehalose and 0.5% sodium azide.
Reconstitution:	Reconstitute with 100 uL sterile water.

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Storage: Aliquot and store at -20C for up to six months after date of receipt. Avoid freeze-thaw cycles.

Expiry Date: 12 months after purchase unopened.

General References:

1. Kleijnen MF, et al. The hPLIC proteins may provide a link between the ubiquitination machinery and the proteasome. Molec. Cell 6: 409-419 (2000).

2. N'Diaye EN, et al. PLIC proteins or ubiquilins regulate autophagy-dependent cell survival during nutrient starvation. EMBO Rep. 10:173-9 (2009).

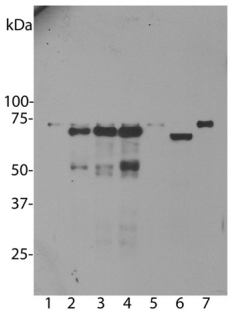
3. Rothenberg C, Srinivasan D, Mah L, Kaushik S, Peterhoff CM, Ugolino J, Fang S, Cuervo AM, Nixon RA, Monteiro MJ. Ubiquilin functions in autophagy and is degraded by chaperone-mediated autophagy. Hum Mol Genet. Aug 15;19 (16): 3219-32. Epub Jun 7 (2010).

4. Deng HX, et al. Mutations in UBQLN2 cause dominant X-linked juvenile and adult-onset ALS and ALS/dementia. Nature. 2011 Aug 21;477(7363):211-5

5. Brettschneider J, et al. Pattern of ubiquilin pathology in ALS and FTLD indicates presence of c9orf72 hexanucleotide expansion. Acta Neuropathol. 2012 Jun;123(6):825-39

6. Renton AE, Majounie E, Waite AA, et al. Hexanucleotide repeat expansion in C9ORF72 is the cause of chromosome 9p21-linked ALS-FTD. Neuron. 2011 Oct 20;72(2):257-68

7. DeJesus-Hernandez M, et al. Expanded GGGGCC hexanucleotide repeat in noncoding region of C9ORF72 causes chromosome 9p-linked FTD and ALS. Neuron. 2011 Oct 20;72(2):245-56

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