



## Anti-UBQLN2 Antibody

**Alternative Names:** ALS15, CHAP1, DSK2, HRIHFB2157, N4BP4, PLIC2, Ubiquilin-2

**Catalogue Number:** AB18-10055-100ug

**Size:** 100 µg

### Background Information

Ubiquilin-2 (UBQLN2) is a 624-amino acid multi-domain adaptor protein and a member of the ubiquilin family of proteins that regulate the degradation of ubiquitinated proteins by the ubiquitin-proteasome system (UPS), autophagy and the endoplasmic reticulum-associated protein degradation (ERAD) pathway. Ubiquilins are characterised by the presence of an N-terminal ubiquitin-like domain and a C-terminal ubiquitin-associated domain. The central portion is highly variable.

UBQLN2 Mediates the proteasomal targeting of misfolded or accumulated proteins for degradation by binding to their polyubiquitin chains, through the ubiquitin-associated domain (UBA) and by interacting with the subunits of the proteasome through the ubiquitin-like domain (ULD). Mutations in UBQLN2 are associated with Amyotrophic Lateral Sclerosis with most ALS-linked mutations localised to the proline-rich repeat (Pxx) region that is unique to ubiquilin-2 and not present in the other members of the ubiquilin protein family.

UBQLN2 has also been shown to bind the ATPase domain of the Hsp70-like Stch protein. Mutations in UBQLN2 are also observed in familial ALS (FALS) cases associated with aberrant TDP-43 inclusions.

### Product Information

<b>Antibody Type:</b>	Polyclonal	<b>Host:</b>	Rabbit
<b>Isotype:</b>	IgG	<b>Species Reactivity:</b>	Human, Mouse
<b>Immunogen:</b>	Partial length recombinant human UBQLN2 from the N-terminal region		
<b>Format:</b>	100 µg in 100 µl PBS containing 0.02% sodium azide.		
<b>Storage Conditions:</b>	6 months: 4°C. Long-term storage: -20°C. Avoid multiple freeze and thaw cycles.		
<b>Applications:</b>	WB WB 1:200-2000.		

### Additional Information

<b>Subcellular location:</b>	Cytoplasm, Cytoplasmic vesicle, Membrane, Nucleus, autophagosome	<b>MW:</b>	66kDa (Intended as a general guide and does not allow for all isoforms and species variations)
<b>Gene ID</b>	29978	<b>Uniprot ID:</b>	Q9UHD9



## References

1. Deng, H. X. et al. Mutations in UBQLN2 cause dominant X-linked juvenile and adult-onset ALS and ALS/dementia. *Nature* 477, 211–215 (2011).

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