

KLHL1 (3A8B3): sc-517225

BACKGROUND

KLHL1 (kelch-like protein 1) is a 748 amino acid protein that is the homolog of the *Drosophila* kelch protein. Localized to the cytoskeleton, KLHL1 contains six kelch repeats and one BTB (POZ) domain. The BTB (broad-complex, tram-track and bric a brac) domain, also known as the POZ (poxvirus and zinc finger) domain, is an N-terminal homodimerization domain that contains multiple copies of kelch repeats and/or C₂H₂-type zinc fingers. KLHL1 is highly expressed in brain where it acts as an Actin-organization protein, possibly playing a role in the modulation of neurite outgrowth. KLHL1 expression has been shown to be downregulated by spinocerebellar ataxia type 8 (SCA8) protein, which then leads to SCA8 neuropathogenesis, a disease that is characterized by limb and truncal ataxia, ataxic dysarthria and horizontal nystagmus.

REFERENCES

1. Nemes, J.P., et al. 2000. The SCA8 transcript is an antisense RNA to a brain-specific transcript encoding a novel actin-binding protein (KLHL1). *Hum. Mol. Genet.* 9: 1543-1551.
2. Melnick, A., et al. 2000. In-depth mutational analysis of the promyelocytic leukemia zinc finger BTB/POZ domain reveals motifs and residues required for biological and transcriptional functions. *Mol. Cell. Biol.* 20: 6550-6567.
3. Adams, J., et al. 2000. The kelch repeat superfamily of proteins: propellers of cell function. *Trends Cell Biol.* 10: 17-24.
4. Prag, S. and Adams, J.C. 2003. Molecular phylogeny of the kelch-repeat superfamily reveals an expansion of BTB/kelch proteins in animals. *BMC Bioinformatics* 4: 42.
5. He, Y., et al. 2006. Targeted deletion of a single Sca8 ataxia locus allele in mice causes abnormal gait, progressive loss of motor coordination, and Purkinje cell dendritic deficits. *J. Neurosci.* 26: 9975-9982.
6. Seng, S., et al. 2006. KLHL1/MRP2 mediates neurite outgrowth in a glycogen synthase kinase 3 β -dependent manner. *Mol. Cell. Biol.* 26: 8371-8384.
7. Jiang, S., et al. 2007. Process elongation of oligodendrocytes is promoted by the Kelch-related protein MRP2/KLHL1. *J. Biol. Chem.* 282: 12319-12329.
8. Aromolaran, K.A., et al. 2007. The Kelch-like protein 1 modulates P/Q-type calcium current density. *Neuroscience* 145: 841-850.
9. Chen, W.L., et al. 2008. SCA8 mRNA expression suggests an antisense regulation of KLHL1 and correlates to SCA8 pathology. *Brain Res.* 1233: 176-184.

CHROMOSOMAL LOCATION

Genetic locus: KLHL1 (human) mapping to 13q21.33.

SOURCE

KLHL1 (3A8B3) is a mouse monoclonal antibody raised against a recombinant protein corresponding to amino acids 202-506 of KLHL1 of human origin.

PRODUCT

Each vial contains 100 μ g IgG₁ in 1.0 ml PBS with < 0.1% sodium azide and 0.1% gelatin.

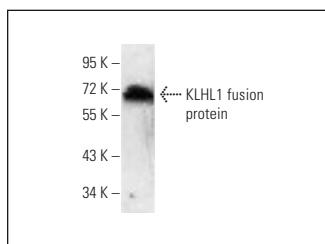
APPLICATIONS

KLHL1 (3A8B3) is recommended for detection of KLHL1 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for KLHL1 siRNA (h): sc-75393, KLHL1 shRNA Plasmid (h): sc-75393-SH and KLHL1 shRNA (h) Lentiviral Particles: sc-75393-V.

Molecular Weight of KLHL1: 83 kDa.

DATA



KLHL1 (3A8B3): sc-517225. Western blot analysis of human recombinant KLHL1 (202-506) fusion protein.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.