

AIPL1 (C-14): sc-46718

BACKGROUND

The inherited blindness associated protein, aryl hydrocarbon receptor interacting protein-like 1 (AIPL1), interacts with the cell cycle regulator protein NUB1. AIPL1 is crucial for protein folding and stabilization, as well as for protein trafficking. It localizes to the nucleus or cytoplasm and is highly expressed in the pineal gland and the retina. In the retina, AIPL1 is expressed in both developing cone and rod photoreceptors, but it is restricted to rod photoreceptors in the adult human retina. Defects in the gene encoding for AIPL1 can cause Leber congenital amaurosis type IV, an early-onset, inherited autosomal recessive disorder that results in childhood blindness.

REFERENCES

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2. van der Spuy, J., et al. 2004. Role of AIP and its homologue the blindness-associated protein AIPL1 in regulating client protein nuclear translocation. *Biochem. Soc. Trans.* 32: 643-645.
3. Dyer, M.A., et al. 2004. Retinal degeneration in AIPL1-deficient mice: a new genetic model of Leber congenital amaurosis. *Brain Res. Mol. Brain Res.* 132: 208-220.
4. Allikmets, R. 2004. Leber congenital amaurosis: a genetic paradigm. *Ophthalmic Genet.* 25: 67-79.
5. Gallon, V.A., et al. 2004. Purification, characterisation and intracellular localisation of aryl hydrocarbon interacting protein-like 1 (AIPL1) and effects of mutations associated with inherited retinal dystrophies. *Biochim. Biophys. Acta* 1690: 141-149.
6. Silva, E., et al. 2004. A missense mutation in GUCY2D acts as a genetic modifier in RPE65-related Leber congenital amaurosis. *Ophthalmic Genet.* 25: 205-217.
7. Liu, X., et al. 2004. AIPL1, the protein that is defective in Leber congenital amaurosis, is essential for the biosynthesis of retinal rod cGMP phosphodiesterase. *Proc. Natl. Acad. Sci. USA* 101: 13903-13908.

CHROMOSOMAL LOCATION

Genetic locus: AIPL1 (human) mapping to 17p13.2; Aipl1 (mouse) mapping to 11 B4.

SOURCE

AIPL1 (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of AIPL1 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-46718 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

AIPL1 (C-14) is recommended for detection of AIPL1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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); may cross-react with AIPL2.

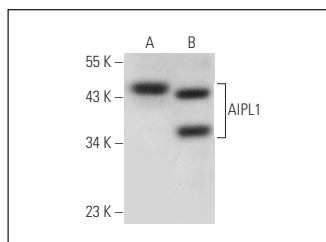
AIPL1 (C-14) is also recommended for detection of AIPL1 in additional species, including equine, bovine and porcine.

Suitable for use as control antibody for AIPL1 siRNA (h): sc-60062, AIPL1 siRNA (m): sc-60063, AIPL1 shRNA Plasmid (h): sc-60062-SH, AIPL1 shRNA Plasmid (m): sc-60063-SH, AIPL1 shRNA (h) Lentiviral Particles: sc-60062-V and AIPL1 shRNA (m) Lentiviral Particles: sc-60063-V.

Molecular Weight of AIPL1: 43 kDa.

Positive Controls: Y79 cell lysate: sc-2240 or ARPE-19 whole cell lysate: sc-364357.

DATA



AIPL1 (C-14): sc-46718. Western blot analysis of AIPL1 expression in Y79 (A) and ARPE-19 (B) whole cell lysates.

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.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.

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Try **AIPL1 (5-RY34): sc-134253**, our highly recommended monoclonal alternative to AIPL1 (C-14).