SANTA CRUZ BIOTECHNOLOGY, INC.

ARH (m): 293T Lysate: sc-124988

BACKGROUND

ARH (autosomal recessive hypercholesterolemia protein), also known as LDLRAP1 (low density lipoprotein receptor adapter protein 1), is a 308 amino acid cytoplasmic protein that contains one PID domain. ARH is an adapter protein required for efficient endocytosis of the LDL receptor (LDLR) from coated pits in polarized cells such as hepatocytes and lymphocytes. To do this, ARH acts to stabilize the interaction between the receptor and the structural components of the pits. While expressed at high levels in kidney, liver and placenta, ARH is expressed at low levels in brain, heart, muscle, colon, spleen, intestine, lung and leukocytes. Defects in the ARH gene are the cause of autosomal recessive hypercholesterolemia, a disorder caused by defective internalization of LDL receptors (LDLR) in the liver. Autosomal recessive hypercholesterolemia has the clinical features of familial hypercholesterolemia (FH), including severely elevated plasma LDL cholesterol, tuberous and tendon xanthomata, and premature atherosclerosis.

REFERENCES

1. Garcia, C.K., et al. 2001. Autosomal recessive hypercholesterolemia caused by mutations in a putative LDL receptor adaptor protein. Science 292: 1394-1398.
2. Online Mendelian Inheritance in Man, OMIM™. 2001. Johns Hopkins University, Baltimore, MD. MIM Number: 605747. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
3. Al-Kateb, H., et al. 2002. Mutation in the ARH gene and a chromosome 13q locus influence cholesterol levels in a new form of digenic-recessive familial hypercholesterolemia. Circ. Res. 90: 951-958.
4. Wilund, K.R., et al. 2002. Molecular mechanisms of autosomal recessive hypercholesterolemia. Hum. Mol. Genet. 11: 3019-3030.
5. He, G., et al. 2002. ARH is a modular adaptor protein that interacts with the LDL receptor, clathrin, and AP-2. J. Biol. Chem. 277: 44044-44049.
6. Mishra, S.K., et al. 2002. The autosomal recessive hypercholesterolemia (ARH) protein interfaces directly with the clathrin-coat machinery. Proc. Natl. Acad. Sci. USA 99: 16099-16104.
7. Mishra, S.K., et al. 2005. Functional dissection of an AP-2 β2 appendage-binding sequence within the autosomal recessive hypercholesterolemia protein. J. Biol. Chem. 280: 19270-19280.
8. Sirinian, M.I., et al. 2005. Adaptor protein ARH is recruited to the plasma membrane by low density lipoprotein (LDL) binding and modulates endocytosis of the LDL/LDL receptor complex in hepatocytes. J. Biol. Chem. 280: 38416-38423.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

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

CHROMOSOMAL LOCATION

Genetic locus: Ldlrap1 (mouse) mapping to 4 D3.

PRODUCT

ARH (m): 293T Lysate represents a lysate of mouse ARH transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

ARH (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive ARH antibodies. Recommended use: 10-20 µl per lane.

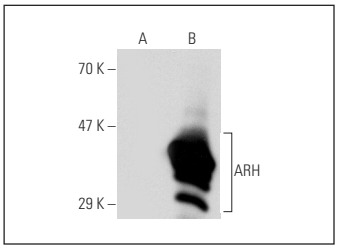
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

ARH (Q-13): sc-100653 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse ARH expression in ARH transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

DATA



ARH (Q-13): sc-100653. Western blot analysis of ARH expression in non-transfected: sc-117752 (**A**) and mouse ARH transfected: sc-124988 (**B**) 293T whole cell lysates.

RESEARCH USE

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