

ABCD1 (H-145): sc-20971

BACKGROUND

The peroxisomal membrane contains several ATP-binding cassette (ABC) transporters, ABCD1-4 that are known to be present in the human peroxisome membrane. All four proteins are ABC half-transporters, which dimerize to form an active transporter. A mutation in the ABCD1 causes X-linked adrenoleukodystrophy (X-ALD), a peroxisomal disorder which affects lipid storage. ABCD2 in mouse is expressed at high levels in the brain and adrenal organs, which are adversely affected in X-ALD. The peroxisomal membrane comprises two quantitatively major proteins, 22 kDa PMP22 and 70 kDa ABCD3. ABCD3 is associated with irregularly shaped vesicles which may be defective peroxisomes or peroxisome precursors. ABCD1 is a 75 kDa protein that localizes to peroxisomes. The genes which encode ABCD1-4 map to human chromosome Xq28, 12q11-q12, 1p22-p21 and 14q24.3, respectively. ABCB7 is a half-transporter involved in the transport of heme from the mitochondria to the cytosol and maps to human chromosome Xq13.1-q13.3.

REFERENCES

- Gartner, J., Kearns, W., Pearson, P. and Valle, D. 1992. Characterization and localization of the human 70 kDa peroxisomal membrane protein (PMP70) gene. (Abstract) Am. J. Hum. Genet. 51: 168.
- Lombard-Platet, G., Savary, S., Sarde, C.O., Mandel, J.L. and Chimini, G. 1996. A close relative of the adrenoleukodystrophy (ALD) gene codes for a peroxisomal protein with a specific expression pattern. Proc. Natl. Acad. Sci. USA 93: 1265-1269.
- Shani, N., Jimenez-Sanchez, G., Steel, G., Dean, M. and Valle, D. 1997. Identification of a fourth half ABC transporter in the human peroxisomal membrane. Hum. Mol. Genet. 6: 1925-1931.
- Moser, H.W. 1997. Adrenoleukodystrophy: phenotype, genetics, pathogenesis and therapy. Brain 120: 1485-1508.
- Savary, S., Troffer-Charlier, N., Gyapay, G., Mattei, M.G. and Chimini, G. 1997. Chromosomal localization of the adrenoleukodystrophy-related gene in man and mice. Eur. J. Hum. Genet. 5: 99-101.
- Holzinger, A., Roscher, A.A., Landgraf, P., Lichtner, P. and Kammerer, S. 1998. Genomic organization and chromosomal localization of the human peroxisomal membrane protein-1-like protein (PXMP1-L) gene encoding a peroxisomal ABC transporter. FEBS Letts. 426: 238-242.
- Shimada, Y., Okuno, S., Kawai, A., Shinomiya, H., Saito, A., Suzuki, M., Omori, Y., Nishino, N., Kanemoto, N., Fujiwara, T., Horie, M. and Takahashi, E. 1998. Cloning and chromosomal mapping of a novel ABC transporter gene (hABC7), a candidate for X-linked sideroblastic anemia with spinocerebellar ataxia. J. Hum. Genet. 43: 115-122.

CHROMOSOMAL LOCATION

Genetic locus: ABCD1 (human) mapping to Xq28; Abcd1 (mouse) mapping to X B.

SOURCE

ABCD1 (H-145) is a rabbit polyclonal antibody raised against amino acids 1-145 of ABCD1 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.

APPLICATIONS

ABCD1 (H-145) is recommended for detection of ABCD1 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).

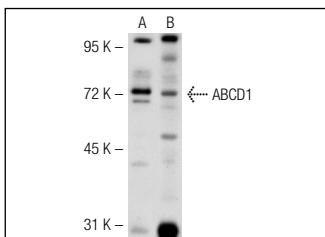
ABCD1 (H-145) is also recommended for detection of ABCD1 in additional species, including porcine.

Suitable for use as control antibody for ABCD1 siRNA (h): sc-41143, ABCD1 siRNA (m): sc-41144, ABCD1 shRNA Plasmid (h): sc-41143-SH, ABCD1 shRNA Plasmid (m): sc-41144-SH, ABCD1 shRNA (h) Lentiviral Particles: sc-41143-V and ABCD1 shRNA (m) Lentiviral Particles: sc-41144-V.

Molecular Weight of ABCD1: 75 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200 or mouse brain extract: sc-2253.

DATA



ABCD1 (H-145): sc-20971. Western blot analysis of ABCD1 expression in 293T whole cell lysate (**A**) and mouse brain tissue extract (**B**).

SELECT PRODUCT CITATIONS

- Hour, T.C., et al. 2009. Downregulation of ABCD1 in human renal cell carcinoma. Int. J. Biol. Markers. 24: 171-178.

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.