

ABCD1 (E-17): sc-31876

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, PMP22 and ABCD3. ABCD3 is associated with irregularly shaped vesicles which may be defective peroxisomes or peroxisome precursors. ABCD4 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. *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.

CHROMOSOMAL LOCATION

Genetic locus: ABCD1 (human) mapping to Xq28; Abcd1 (mouse) mapping to X A7.3.

SOURCE

ABCD1 (E-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus 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.

Blocking peptide available for competition studies, sc-31876 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

ABCD1 (E-17) 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), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 (E-17) is also recommended for detection of ABCD1 in additional species, including equine, canine, bovine and 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.

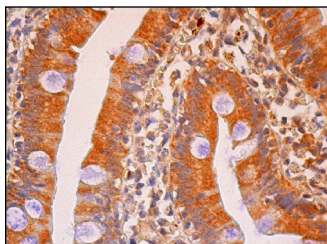
Molecular Weight of ABCD1: 75 kDa.

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

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



ABCD1 (E-17): sc-31876. Immunoperoxidase staining of formalin fixed, paraffin-embedded human small intestine tissue showing cytoplasmic staining of glandular cells.

SELECT PRODUCT CITATIONS

- Kao, Y.T., Hsu, W.C., Hu, H.T., Hsu, S.H., Lin, C.S., Chiu, C.C., Lu, C.Y., Hour, T.C., Pu, Y.S. and Huang, A.M. 2014. Involvement of p38 mitogen-activated protein kinase in acquired gemcitabine-resistant human urothelial carcinoma sublines. *Kaohsiung J. Med. Sci.* 30: 323-330.

RESEARCH USE

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