

Fibrocystin (H-300): sc-99139

BACKGROUND

Fibrocystin is a type I membrane protein that undergoes regulated proteolysis. Many proteolytic cleavages occur on the ectodomain whereas at least one cleavage occurs on the cytoplasmic portion of Fibrocystin. The latter generates a C-terminal intracellular fragment that localizes to the nucleus. This proteolysis requires activation of protein kinase C (PKC) and release of intracellular calcium. Fibrocystin is expressed in the cilia of the bile duct epithelium and leads to abnormalities in the rubric of the ductal plate malformation. The intracellular C-terminus of Fibrocystin interacts with calcium modulating cyclophilin ligand (CAML), a protein implicated in calcium signaling. Fibrocystin may participate in the mediation of intracellular calcium in the distal nephron in a manner similar to PKD1 and PKD2. Mutations in the PKHD1 gene, which encodes Fibrocystin, result in autosomal recessive polycystic kidney disease (ARPKD), a severe form of polycystic kidney disease characterized by enlarged kidneys and congenital hepatic fibrosis.

REFERENCES

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- Peters, D.J., et al. 2005. From gene to disease; PKHD1 and recessive polycystic kidney disease. *Ned. Tijdschr. Geneesk.* 149: 463-466.
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CHROMOSOMAL LOCATION

Genetic locus: PKHD1 (human) mapping to 6p12.3; Pkhd1 (mouse) mapping to 1 A3.

SOURCE

Fibrocystin (H-300) is a rabbit polyclonal antibody raised against amino acids 181-480 mapping within an N-terminal extracellular domain of Fibrocystin 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

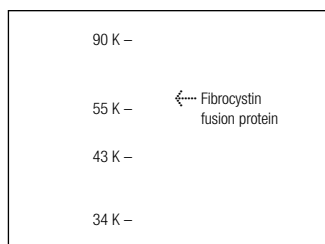
Fibrocystin (H-300) is recommended for detection of Fibrocystin isoforms 1 and 2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Fibrocystin (H-300) is also recommended for detection of Fibrocystin isoforms 1 and 2 in additional species, including equine, canine and bovine.

Suitable for use as control antibody for Fibrocystin siRNA (h): sc-60637, Fibrocystin siRNA (m): sc-60638, Fibrocystin shRNA Plasmid (h): sc-60637-SH, Fibrocystin shRNA Plasmid (m): sc-60638-SH, Fibrocystin shRNA (h) Lentiviral Particles: sc-60637-V and Fibrocystin shRNA (m) Lentiviral Particles: sc-60638-V.

Molecular Weight of Fibrocystin: 447 kDa.

DATA



Fibrocystin (H-300): sc-99139. Western blot analysis of human recombinant Fibrocystin fusion protein.

SELECT PRODUCT CITATIONS

- Sun, L., et al. 2010. Regulation of cell proliferation and apoptosis through fibrocystin-prosaposin interaction. *Arch. Biochem. Biophys.* 502: 130-136.

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.