

Fibrocystin L (W-13): sc-87744

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. 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. A related protein, Fibrocystin L, also designated polycystic kidney and hepatic disease 1-like protein 1 or PKHD1L1, shares 41% similarity with Fibrocystin in the extracellular domain, but is not associated with ARPKD. Fibrocystin L is a large receptor protein with a signal peptide, a single transmembrane domain and a short cytoplasmic tail. It is ubiquitously expressed at low levels, with higher expression in spleen and thymus as well as in activated T cells and B lymphoblasts, suggesting a role for Fibrocystin L in the immune response.

REFERENCES

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RESEARCH USE

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

CHROMOSOMAL LOCATION

Genetic locus: PKHD1L1 (human) mapping to 8q23.1; Pkhd1l1 (mouse) mapping to 15 B3.2.

SOURCE

Fibrocystin L (W-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of Fibrocystin L 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-87744 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Fibrocystin L (W-13) is recommended for detection of Fibrocystin L 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Fibrocystin L (W-13) is also recommended for detection of Fibrocystin L in additional species, including canine, bovine and porcine.

Suitable for use as control antibody for Fibrocystin L siRNA (h): sc-77480, Fibrocystin L siRNA (m): sc-145175, Fibrocystin L shRNA Plasmid (h): sc-77480-SH, Fibrocystin L shRNA Plasmid (m): sc-145175-SH, Fibrocystin L shRNA (h) Lentiviral Particles: sc-77480-V and Fibrocystin L shRNA (m) Lentiviral Particles: sc-145175-V.

Molecular Weight of Fibrocystin L: 466 kDa.

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.

STORAGE

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

PROTOCOLS

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