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**


**CHROMOSOMAL LOCATION**

Genetic locus: PKHDL1 (human) mapping to 8q23.1; Pkhdl11 (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).


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, dilution range 1:100-1:1000), Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2035, TBS Blotto A Blocking Reagent or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2048 (dilution range 1:100-1:1000), 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-2020 (dilution range 1:200, dilution range 1:100-1:1000), immunofluorescence: use donkey anti-goat IgG-HRP: sc-2048 (dilution range 1:100-1:1000), donkey anti-goat IgG-TR: sc-2024 (dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000). 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.

**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.