

# Fibrocystin (C-20): sc-49671

## 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

1. Masyuk, T.V., et al. 2003. Defects in cholangiocyte fibrocystin expression and ciliary structure in the PCK rat. *Gastroenterology* 125: 1303-1310.
2. Ward C.J., et al. 2003. Cellular and subcellular localization of the ARPKD protein; fibrocystin is expressed on primary cilia. *Hum. Mol. Genet.* 12: 2703-2710.
3. Bergmann, C., et al. 2005. Algorithm for efficient PKHD1 mutation screening in autosomal recessive polycystic kidney disease (ARPKD). *Hum. Mutat.* 25: 225-231.
4. Bergmann, C., et al. 2005. Clinical consequences of PKHD1 mutations in 164 patients with autosomal-recessive polycystic kidney disease (ARPKD). *Kidney Int.* 67: 829-848.
5. Losekoot, M., et al. 2005. Analysis of missense variants in the PKHD1 gene in patients with autosomal recessive polycystic kidney disease (ARPKD). *Hum. Genet.* 118: 185-206.
6. Mai, W., et al. 2005. Inhibition of PKHD1 impairs tubulomorphogenesis of cultured IMCD cells. *Mol. Biol. Cell* 16: 4398-4409.
7. Nagano, J., et al. 2005. Fibrocystin interacts with CAML, a protein involved in Ca<sup>2+</sup> signaling. *Biochem. Biophys. Res. Commun.* 338: 880-889.

## CHROMOSOMAL LOCATION

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

## SOURCE

Fibrocystin (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus 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.

Blocking peptide available for competition studies, sc-49671 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

Fibrocystin (C-20) is recommended for detection of Fibrocystin isoform 1 of human and, to a lesser extent, mouse and rat 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).

Fibrocystin (C-20) is also recommended for detection of Fibrocystin isoform 1 in additional species, including equine, canine and bovine.

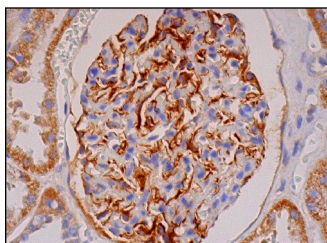
Suitable for use as control antibody for Fibrocystin siRNA (h): sc-60637, Fibrocystin siRNA (m): sc-60638, Fibrocystin shRNA Plasmid (h): sc-60637-SH, Fibrocystin shRNA Plasmid (m): sc-60638-SH, Fibrocystin shRNA (h) Lentiviral Particles: sc-60637-V and Fibrocystin shRNA (m) Lentiviral Particles: sc-60638-V.

Molecular Weight of Fibrocystin: 447 kDa.

## 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



Fibrocystin (C-20): sc-49671. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing membrane staining of cells in glomeruli and cytoplasmic staining of cells in tubules.

## 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.