



PKHD1 (Polycystic Kidney and Hepatic Disease 1, ARPKD, DKFZp686C01112, Fibrocystin, FCYT, FLJ46150, Polyductin, Tigmin, TIGM1)

Catalog number

P4210-70A

Supplier

United States Biological

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 later 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.

Applications

Suitable for use in Peptide ELISA. Other applications not tested.

Recommended Dilution

Peptide ELISA: 1:100-1:2000

Optimal dilutions to be determined by the researcher.

Storage and Stability

May be stored at 4°C for short-term only. Aliquot to avoid repeated freezing and thawing. Store at -20°C. Aliquots are stable for 12 months after receipt. For maximum recovery of product, centrifuge the original vial after thawing and prior to removing the cap.

Immunogen

Synthetic peptide derived from the human PKHD1 protein (within residues 100-200). Species sequence homology: mouse (93%). Cellular Localization: Membrane; single-pass type I membrane.

Formulation

Supplied as a liquid in PBS, pH 7.2, 0.05% sodium azide, 30% glycerol.

Purity

Purified by immunoaffinity chromatography.

Specificity

Recognizes human PKHD1

Product Type



Pab

Source

human

Isotype

IgG

Grade

Affinity Purified

Applications

E

Crossreactivity

Hu

Storage

-20°C