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

**Catalog number**

P4210-70B

**Supplier**

United States Biological

PKHD1 protein (polycystic kidney and hepatic disease 1 protein) is encoded by PKHD1 gene whose mutations have been established as underlying cause of autosomal recessive polycystic kidney disease (ARPKD), a rare genetic disorder characterized mainly by cystic kidneys in the developing fetus in utero and post-natal pulmonary insufficiency as well as renal manifestations, abnormal biliary development with dilated bile ductules and peribiliary fibrosis, collectively termed congenital hepatic fibrosis (CHF). PKHD1 is a type I membrane protein with a large extracellular N-terminal domain, a single transmembrane segment, and a short cytoplasmic C-terminus. PKHD1 is a large protein with predicted molecular weight of 447 kD and is predominantly expressed in kidneys (the cortical and medullary collecting ducts), pancreas, liver etc. and in addition to other locations in the cells, it is localized on primary cilium, hair-like organelle present on cellular surfaces. PKHD1 is essential for correct bipolar cell division through the regulation of centrosome duplication and mitotic spindle assembly. Moreover, it has been suggested to act as a receptor that plays a role in collecting duct/biliary differentiation and lack of PKHD1 results in stunted primary cilia which have been proposed to underlie the pathogenesis of ARPKD.

**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 corresponding to human PKHD1 (within residues 450-550). Localization: Membrane; single-pass type I membrane. Species sequence homology: mouse (88%)

**Formulation**

Supplied as a liquid in PBS, pH 7.2, 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