

## PKHD1 Antibody

Catalog No: #48398

Package Size: #48398-1 50ul #48398-2 100ul

Orders: order@signalwayantibody.com

Support: tech@signalwayantibody.com

## Description

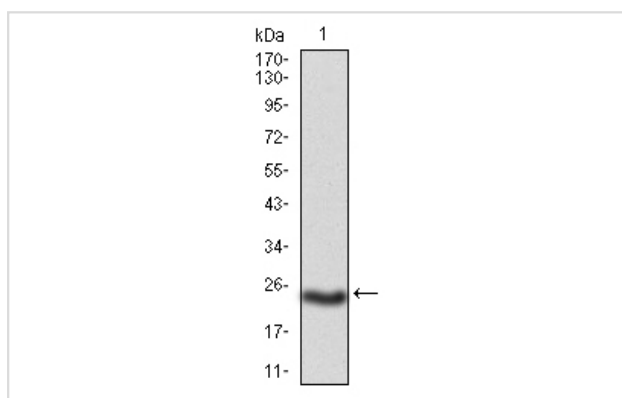
Product Name	PKHD1 Antibody
Host Species	Mouse
Clonality	Monoclonal
Clone No.	A1-G12
Purification	ProA affinity purified
Applications	WB, ICC, IHC, FC
Species Reactivity	Hu
Immunogen Description	Recombinant protein
Other Names	ARPKD antibody FCYT antibody Fibrocystin antibody FPC antibody PKHD1 antibody PKHD1_HUMAN antibody Polycystic kidney and hepatic disease 1 protein antibody Polyductin antibody TIGM1 antibody Tigmin antibody
Accession No.	Swiss-Prot#:P08F94
Uniprot	P08F94
GeneID	5314;
Calculated MW	445 kDa
Formulation	1*TBS (pH7.4), 1%BSA, 40%Glycerol. Preservative: 0.05% Sodium Azide.
Storage	Store at -20°C

## Application Details

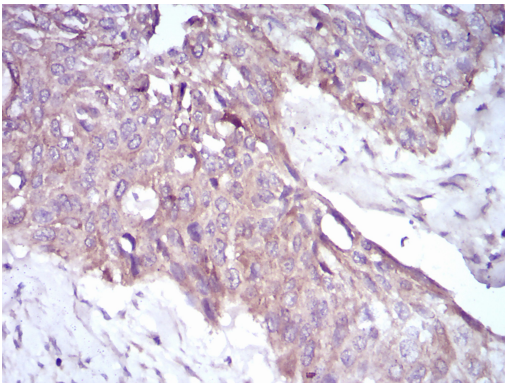
WB: 1:500-1:2,000 IHC: 1:50-1:200

ICC: 1:50-1:200 FC: 1:50-1:100

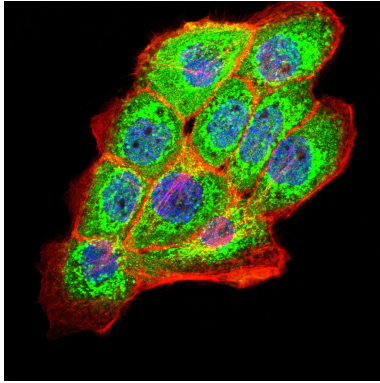
## Images



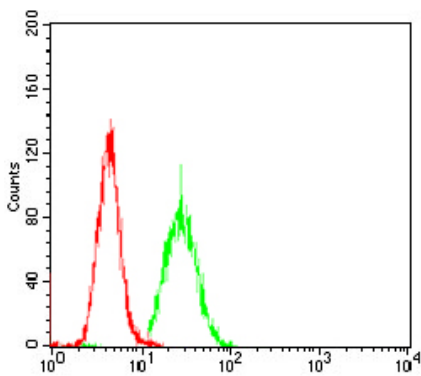
Western blot analysis of PKHD1 on mouse PKHD1 recombinant protein using anti-PKHD1 antibody at 1/1,000 dilution.



Immunohistochemical analysis of paraffin-embedded human esophageal cancer tissue using anti-PKHD1 antibody. Counter stained with hematoxylin.



ICC staining PKHD1 (green) and Actin filaments (red) in A431 cells. The nuclear counter stain is DAPI (blue). Cells were fixed in paraformaldehyde, permeabilised with 0.25% Triton X100/PBS.



Flow cytometric analysis of HeLa cells with PKHD1 antibody at 1/100 dilution (green) compared with an unlabelled control (cells without incubation with primary antibody; red).

## Background

May be required for correct bipolar cell division through the regulation of centrosome duplication and mitotic spindle assembly. May be a receptor protein that acts in collecting-duct and biliary differentiation. Defects in PKHD1 are the cause of polycystic kidney disease autosomal recessive (ARPKD). ARPKD is a severe form of polycystic kidney disease affecting the kidneys and the hepatic biliary tract. The clinical spectrum is widely variable, with most cases presenting during infancy. The fetal phenotypic features classically include enlarged and echogenic kidneys, as well as oligohydramnios secondary to a poor urine output. Up to 50% of the affected neonates die shortly after birth, as a result of severe pulmonary hypoplasia and secondary respiratory insufficiency. In the subset that survives the perinatal period, morbidity and mortality are mainly related to severe systemic hypertension, renal insufficiency, and portal hypertension due to portal-tract fibrosis.

## References

Note: This product is for in vitro research use only