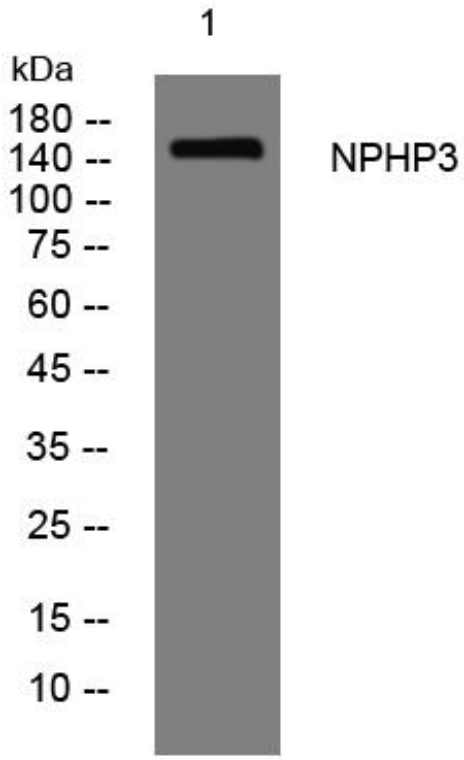
NPHP3 mouse mAb

Catalog No	BYmab-11324
Isotype	IgG
Reactivity	Human; Mouse
Applications	WB
Gene Name	NPHP3 KIAA2000
Protein Name	NPHP3
Immunogen	Synthesized peptide derived from human NPHP3 AA range: 1092-1142
Specificity	This antibody detects endogenous levels of NPHP3 at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	Cell projection, cilium . Localization to cilium is mediated via interaction with UNC119 and UNC119B, which bind to the myristoyl moiety of the N-terminus.
Tissue Specificity	Widely expressed at low level. Expressed in heart, placenta, liver, skeletal muscle, kidney and pancreas. Expressed at very low level in brain and lung.
Function	alternative products:Additional isoforms seem to exist,disease:Defects in NPHP3 are a cause of renal-hepatic-pancreatic dysplasia (RHPD) [MIM:208540]. RHPD is an autosomal recessive disorder with variable expression, and patients surviving the neonatal period progress to renal and hepatic failure which can be treated successfully with combined liver-kidney transplantation.,disease:Defects in NPHP3 are the cause of nephronophthisis type 3 (NPHP3) [MIM:604387]; also known as adolescent nephronophthisis. NPHP3 is a autosomal recessive disorder resulting in end-stage renal disease. It is characterized by polyuria, polydipsia, anemia. Onset of terminal renal failure occurr significantly later (median age, 19 years) than in juvenile nephronophthisis. Renal pathology is characterized by alterations of tubular basement membranes, tubular atrophy and dilatation,

	sclerosing tubulointerstitial neph
Background	This gene encodes a protein containing a coiled-coil (CC) domain, a tubulin-tyrosine ligase (TTL) domain, and a tetratrico peptide repeat (TPR) domain. The encoded protein interacts with nephrocystin, it is required for normal ciliary development, and it functions in renal tubular development. Mutations in this gene are associated with nephronophthisis type 3, and also with renal-hepatic-pancreatic dysplasia, and Meckel syndrome type 7. Naturally occurring read-through transcripts exist between this gene and the downstream ACAD11 (acyl-CoA dehydrogenase family, member 11) gene. [provided by RefSeq, Feb 2011],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using NPHP3 mouse mAb