NPHP3 Rabbit pAb

CatalogNo: YT6689

Key Features

Host Species	Reactivity	Applications
• Rabbit	• Human,Mouse	• WB

MW	Isotype
• 146kD (Calculated)	• IgG

Recommended Dilution Ratios

WB 1:500-2000

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Basic Information

Clonality	Polyclonal

Immunogen Information

Immunogen	Synthesized peptide derived from human NPHP3 AA range: 1092-1142
Specificity	This antibody detects endogenous levels of NPHP3 at Human/Mouse

Target Information

Gene name	NPHP3 KIAA2000

Protein Name NPHP3

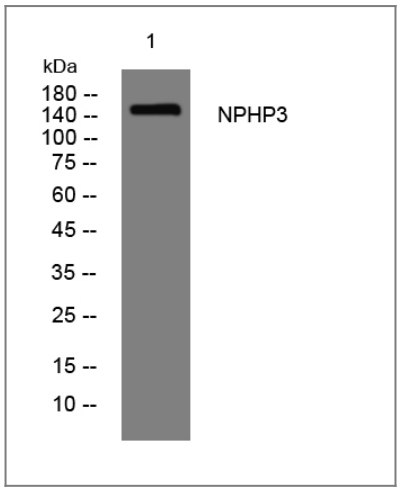
Organism	Gene ID	UniProt ID
Human	27031;	Q7Z494;
Mouse	74025;	Q7TNH6;

Cellular Localization 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 nephropathy, and renal cyst development predominantly at the corticomedullary junction.,Function:May participate in mechanosensation in the primary cilium of kidney cells.,similarity:Belongs to the acyl-CoA dehydrogenase family.,similarity:Contains 11 TPR repeats.,subunit:Interacts with NPHP1.,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.,

Validation Data



Western blot analysis of lysates from Hela cells, primary antibody was diluted at 1:1000, 4°over night

Contact information

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