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Rabbit Anti-NPC2 antibody

SL11736R

Product Name:	NPC2
Chinese Name:	尼曼匹克C2前体蛋白抗体
Alias:	EDDM1; Niemann Pick C2; Epididymal protein 1; Epididymal secretory protein; Epididymal secretory protein E1; HE1; Human epididymis-specific protein 1; Niemann-Pick disease type C2; Niemann-Pick disease type C2 protein; NPC2; NPC2_HUMAN; Tissue specific secretory protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	15kDa
Cellular localization:	cytoplasmicSecretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Niemann Pick C2:20-80/151
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	This gene encodes a protein containing a lipid recognition domain. The encoded protein may function in regulating the transport of cholesterol through the late endosomal/lysosomal system. Mutations in this gene have been associated with Niemann-Pick disease, type C2 and frontal lobe atrophy. [provided by RefSeq, Jul 2008]

Function:
May be involved in the regulation of the lipid composition of sperm membranes during the maturation in the epididymis.

Subunit:
Interacts with NUS1/NgBR, the interaction stabilizes NCP2 and regulates cholesterol trafficking. Interacts with DHDDS. Interacts with NPC1 (via the second lumenal domain) in a cholestrol-dependent manner (By similarity). Interacts with NEDD4L (via C2 domain) (By similarity). Interacts with NPC1L1.

Subcellular Location:
Secreted. Endoplasmic reticulum. Lysosome.

Tissue Specificity:
Epididymis.

DISEASE:
Defects in NPC2 are the cause of Niemann-Pick disease type C2 (NPDC2) [MIM:607625]. A lysosomal storage disorder that affects the viscera and the central nervous system. It is due to defective intracellular processing and transport of low-density lipoprotein derived cholesterol. It causes accumulation of cholesterol in lysosomes, with delayed induction of cholesterol homeostatic reactions. Niemann-Pick disease type C2 has a highly variable clinical phenotype. Clinical features include variable hepatosplenomegaly and severe progressive neurological dysfunction such as ataxia, dystonia and dementia. The age of onset can vary from infancy to late adulthood.

Similarity:
Belongs to the NPC2 family.

SWISS:
P61916

Gene ID:
10577

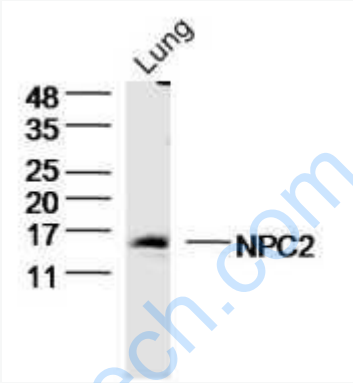
Database links:

Entrez Gene: 10577 Human

Omim: 601015 Human

SwissProt: P61916 Human

SwissProt: P61918 Monkey

	Unigene: 433222 Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: Lung (Mouse) Lysate at 40 ug Primary: Anti- NPC2 (SL11736R)at 1/300 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 15kD Observed band size: 15kD