

## Allgemeine Informationen

<b>Katalog-Nr.:</b> 13926-1-AP	<b>GenBank-Zugangsnummer:</b> BC063302	<b>Reinigungsmethode:</b> Antigen-Affinitätsreinigung
<b>Größe:</b> 150ul , Konzentration: 600 µg/ml von Nanodrop;	<b>GeneID (NCBI):</b> 4864	<b>Empfohlene Verdünnungen:</b> WB 1:500-1:2000 IHC 1:50-1:500 IF 1:50-1:500
<b>Wirt:</b> Kaninchen	<b>Vollständiger Name:</b> Niemann-Pick disease, type C1	
<b>Isotyp:</b> IgG	<b>Berechnete Masse:</b> 142 kDa	
<b>Immunogen Katalognummer:</b> AG4946	<b>Beobachtete Masse:</b> 160-200 kDa	

## Anwendungen

### Geprüfte Anwendungen:

IF, IHC, WB, ELISA

### In Publikationen genannte Anwendungen:

IHC, WB

### Getestete Reaktivität:

Human, Maus

### Zitierte Arten:

Hausschwein, Human, Maus

**Hinweis-IHC: Antigenmaskierung mit TE-Puffer pH 9,0 empfohlen. (\*) Wahlweise kann die Antigenmaskierung auch mit Citratpuffer pH 6,0 erfolgen.**

### Positivkontrollen:

**WB** : ungekochte HEK-293-Zellen, HeLa-Zellen, HepG2-Zellen, ungekochtes Maushirngewebe

**IHC** : humanes Leberkarzinomgewebe, humanes Hirngewebe, humanes Plazenta-Gewebe

**IF** : HepG2-Zellen,

## Hintergrundinformationen

Niemann-Pick Type C (NPC) disease is a lysosomal storage disorder characterized by accumulation of unesterified cholesterol and other lipids in the endolysosomal system. NPC disease results from a defect in either of two distinct cholesterol-binding proteins: a transmembrane protein, NPC1, and a small soluble protein, NPC2. NPC1 or NPC2 deficiency models showed that the functions of these two proteins within lysosomes are linked closely. NPC1 is a typical transmembrane protein and contains a number of modification sites for glycosylation. Defects in NPC1 are the cause of Niemann-Pick disease type C1 which exhibits highly variable clinical phenotype. Moreover, NPC1 may play a role in vesicular trafficking in glia, a process that may be crucial for maintaining the structural and functional integrity of nerve terminals.

## Bemerkenswerte Veröffentlichungen

Verfasser	Pubmed ID	Journal	Anwendung
Junfang Lyu	28923401	Cancer Lett	WB
Guoli Li	34047913	Sci China Life Sci	WB
Jian Xiao	31144242	Sci China Life Sci	

## Lagerung

### Lagerungsbedingungen:

Bei -20°C lagern. Nach dem Versand ein Jahr lang stabil

### Lagerungspuffer:

PBS mit 0.02% Natriumazid und 50% Glycerin pH 7.3.

Aliquotieren ist nicht notwendig bei -20°C Lagerung

**\*\*\* 20ul-Größen enthalten 0.1% BSA**

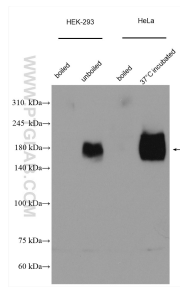
For technical support and original validation data for this product please contact:

T: 1 (888) 4PTGLAB (1-888-478-4522) (toll free in USA), or 1(312) 455-8498 (outside USA)

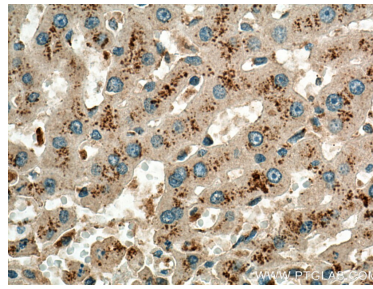
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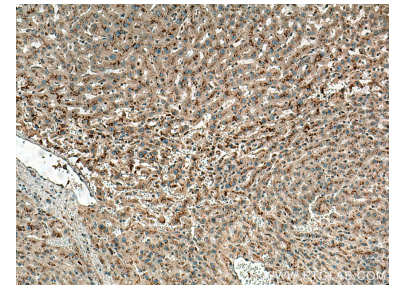
## Ausgewählte Validierungsdaten



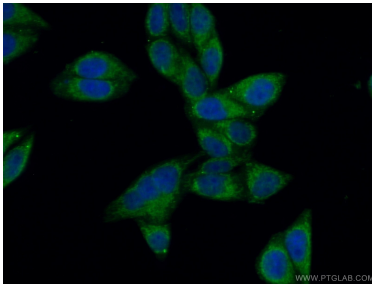
Various lysates were subjected to SDS PAGE followed by western blot with 13926-1-AP (NPC1 antibody) at dilution of 1:1000 incubated at room temperature for 1.5 hours.



Immunohistochemical analysis of paraffin-embedded human liver cancer tissue slide using 13926-1-AP (NPC1 antibody) at dilution of 1:200 (under 40x lens). Heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0).



Immunohistochemical analysis of paraffin-embedded human liver cancer tissue slide using 13926-1-AP (NPC1 antibody) at dilution of 1:200 (under 10x lens). Heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0).



Immunofluorescent analysis of (10% Formaldehyde) fixed HepG2 cells using 13926-1-AP (NPC1 antibody) at dilution of 1:50 and Alexa Fluor 488-conjugated AffiniPure Goat Anti-Rabbit IgG(H+L).