

Allgemeine Informationen

Katalog-Nr.: 30006-1-AP	GenBank-Zugangsnummer: NM_000203	Reinigungsmethode: Antigen-Affinitätsreinigung
Größe: 150ul, Konzentration: 300 µg/ml von Nanodrop;	GeneID (NCBI): 3425	Empfohlene Verdünnungen: WB 1:500-1:3000 IHC 1:50-1:500
Wirt: Kaninchen	Vollständiger Name: iduronidase, alpha-L-	
Isotyp: IgG	Berechnete Masse: 73 kDa	
Immunogen Katalognummer: AG30658	Beobachtete Masse: 73 kDa	

Anwendungen

Geprüfte Anwendungen: IHC, WB, ELISA	Positivkontrollen:
Getestete Reaktivität: Human, Maus	WB : A549-Zellen, HEK-293-Zellen, LNCaP-Zellen, Maushirngewebe, Rattenhirngewebe
Hinweis-IHC: Antigendemaskierung mit TE-Puffer pH 9,0 empfohlen. (*) Wahlweise kann die Antigendemaskierung auch mit Citratpuffer pH 6,0 erfolgen.	IHC : Mausnierengewebe,

Hintergrundinformationen

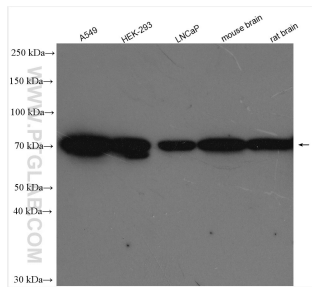
Iduronidase (L-iduronidase, alpha-L-iduronidase, laronidase) is an enzyme with the systematic name glycosaminoglycan alpha-L-iduronohydrolase. This enzyme catalyzes the hydrolysis of unsulfated alpha-L-iduronosidic linkages in dermatan sulfate. It is a glycoprotein enzyme found in the lysosomes of cells. It is involved in the degeneration of glycosaminoglycans such as dermatan sulfate and heparan sulfate. The enzyme acts by hydrolyzing the terminal alpha-L-iduronic acid residues of these molecules, degrading them (PMID: 4993544,30407). A deficiency in the IDUA protein is associated with mucopolysaccharidoses (MPS). MPS, a type of lysosomal storage disease, is typed I through VII. In this syndrome, glycosaminoglycans accumulate in the lysosomes and cause substantial disease in many different tissues of the body. IDUA mutations result in the MPS 1 phenotype, which is inherited in an autosomal recessive fashion. The defective alpha-L-iduronidase results in an accumulation of heparan and dermatan sulfate within phagocytes, endothelium, smooth muscle cells, neurons, and fibroblasts. Prenatal diagnosis of this enzyme deficiency is possible (PMID:8242073).

Lagerung

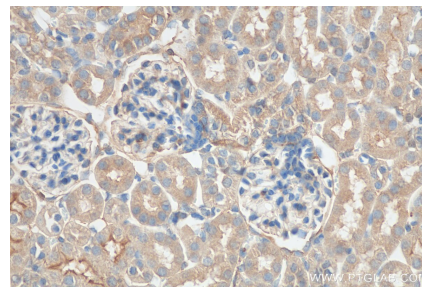
Lagerungsbedingungen:
Bei -20°C lagern. Nach dem Versand ein Jahr lang stabil
Lagerungspuffer:
PBS mit 0.02% Natriumazid und 50% Glycerin pH 7.3.
Aliquotieren ist nicht notwendig bei -20°C Lagerung

*** 20ul-Größen enthalten 0.1% BSA

Ausgewählte Validierungsdaten



Various lysates were subjected to SDS PAGE followed by western blot with 30006-1-AP (IDUA antibody) at dilution of 1:1500 incubated at room temperature for 1.5 hours.



Immunohistochemical analysis of paraffin-embedded mouse kidney tissue slide using 30006-1-AP (IDUA antibody) at dilution of 1:200 (under 40x lens). Heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0).