

ANAG rabbit pAb

Catalog No :	YT6646
Reactivity :	Human
Applications :	WB
Target :	ANAG
Fields :	>>Glycosaminoglycan degradation;>>Metabolic pathways;>>Lysosome
Gene Name :	NAGLU UFHSD1
Protein Name :	ANAG
Human Gene Id :	4669
Human Swiss Prot No :	P54802
Immunogen :	Synthesized peptide derived from human ANAG AA range: 146-196
Specificity :	This antibody detects endogenous levels of ANAG at Human
Formulation :	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source :	Polyclonal, Rabbit,IgG
Dilution :	WB 1[?]500-2000
Purification :	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Concentration :	1 mg/ml
Storage Stability :	-15°C to -25°C/1 year(Do not lower than -25°C)
Molecularweight :	82kD

Background :

This gene encodes an enzyme that degrades heparan sulfate by hydrolysis of terminal N-acetyl-D-glucosamine residues in N-acetyl-alpha-D-glucosaminides. Defects in this gene are the cause of mucopolysaccharidosis type IIIB (MPS-IIIB), also known as Sanfilippo syndrome B. This disease is characterized by the lysosomal accumulation and urinary excretion of heparan sulfate. [provided by RefSeq, Jul 2008],

Function :

catalytic activity:Hydrolysis of terminal non-reducing N-acetyl-D-glucosamine residues in N-acetyl-alpha-D-glucosaminides.,caution:A MPS3B mutation at position 100 was erroneously reported (PubMed:9950362) as an amino acid change from Arg to His. The right amino acid change is from His to Arg.,disease:Defects in NAGLU are the cause of mucopolysaccharidosis type 3B (MPS3B) [MIM:252920]; also known as Sanfilippo syndrome B. MPS3B is a form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life.,functio

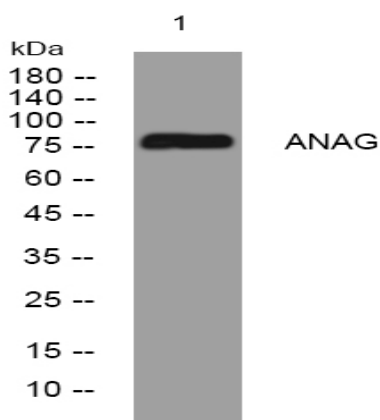
Subcellular Location :

Lysosome.

Expression :

Liver, ovary, peripheral blood leukocytes, testis, prostate, spleen, colon, lung, placenta and kidney.

Products Images



Western blot analysis of lysates from A549 cells, primary antibody was diluted at 1:1000, 4° over night