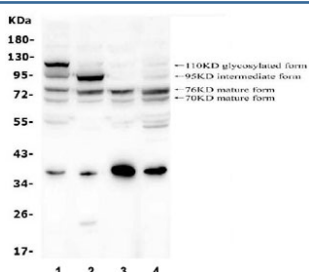


GAA Antibody / Glucosidase alpha acid (R32608)

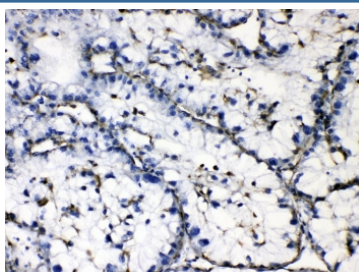
Catalog No.	Formulation	Size
R32608	0.5mg/ml if reconstituted with 0.2ml sterile DI water	100 ug

Bulk quote request

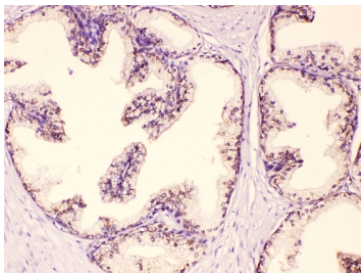
Availability	1-3 business days
Species Reactivity	Human
Format	Antigen affinity purified
Host	Rabbit
Clonality	Polyclonal (rabbit origin)
Isotype	Rabbit IgG
Purity	Antigen affinity
Buffer	Lyophilized from 1X PBS with 2.5% BSA and 0.025% sodium azide
UniProt	P10253
Localization	Cytoplasmic, membranous
Applications	Western Blot : 0.5-1ug/ml Immunohistochemistry (FFPE) : 1-2ug/ml
Limitations	This GAA antibody is available for research use only.



Western blot testing of human 1) A549, 2) HepG2, 3) HEK293 and 4) PC-3 cell lysate with GAA antibody at 0.5ug/ml. Expected molecular weight ~110 kDa (precursor), ~95 kDa (intermediate), ~76 and 70 kDa (lysosomal forms).



IHC testing of FFPE human liver cancer tissue with GAA antibody at 1ug/ml. HIER: steam section in pH6 citrate buffer for 20 min.



IHC testing of FFPE human prostate cancer tissue with GAA antibody at 1ug/ml. HIER: steam section in pH6 citrate buffer for 20 min.

Description

Lysosomal alpha-glucosidase is an enzyme that in humans is encoded by the GAA gene. This gene encodes lysosomal alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. The encoded preproprotein is proteolytically processed to generate multiple intermediate forms and the mature form of the enzyme. Defects in this gene are the cause of glycogen storage disease II, also known as Pompe's disease, which is an autosomal recessive disorder with a broad clinical spectrum. Alternative splicing results in multiple transcript variants.

Application Notes

Optimal dilution of the GAA antibody should be determined by the researcher.

Immunogen

Amino acids 494-527 (TALAWWEDMVAEFHDQVPFDGMWIDMNEPSNFIR-human) were used as the immunogen for the GAA antibody.

Storage

After reconstitution, the GAA antibody can be stored for up to one month at 4oC. For long-term, aliquot and store at -20oC. Avoid repeated freezing and thawing.

References (1)