



Anti-GAA (full length) polyclonal antibody (CPBT-35311RH)

This product is for research use only and is not intended for diagnostic use.

PRODUCT INFORMATION

Product Overview	Rabbit Polyclonal antibody to Human GAA.
Antigen Description	This gene encodes acid alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. Different forms of acid alpha-glucosidase are obtained by proteolytic processing. Defects in this gene are the cause of glycogen storage disease II, also known as Pompe disease, which is an autosomal recessive disorder with a broad clinical spectrum. Three transcript variants encoding the same protein have been found for this gene.
Immunogen	Full length protein, corresponding to amino acids 1-952 of Human GAA.
Isotype	IgG
Source/Host	Rabbit
Species Reactivity	Mouse, Human
Purification	Protein A purified
Conjugate	Unconjugated
Applications	WB
Sequence Similarities	Belongs to the glycosyl hydrolase 31 family. Contains 1 P-type (trefoil) domain.
Cellular Localization	Lysosome. Lysosome membrane.
Format	Liquid
Size	100 µg
Buffer	Preservative: None Constituents: 1X PBS, pH 7.2

Preservative	None
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.

GENE INFORMATION

Gene Name	GAA glucosidase, alpha: acid [Homo sapiens]
Official Symbol	GAA
Synonyms	GAA; glucosidase, alpha; acid; lysosomal alpha-glucosidase; glycogen storage disease type II; Pompe disease; 70 kDa lysosomal alpha-glucosidase; Acid alpha glucosidase; Acid maltase; Aglucosidase alfa; Alpha glucosidase; GAA; Glucosidase alpha acid (Pompe disease glycogen storage disease type II); Glucosidase alpha acid; Glucosidase alpha; LYAG; LYAG_HUMAN; Lysosomal alpha glucosidase; acid maltase; aglucosidase alfa; LYAG;
Entrez Gene ID	2548
Protein Refseq	NP_000143
UniProt ID	P10253
Chromosome Location	17q25.2-q25.3
Pathway	Galactose metabolism, organism-specific biosystem; Galactose metabolism, conserved biosystem; Lysosome, organism-specific biosystem; Lysosome, conserved biosystem; Metabolic pathways, organism-specific biosystem; Notch-mediated HES/HEY network, organism-specific biosystem; Starch and sucrose metabolism, organism-specific biosystem; Starch and sucrose metabolism, conserved biosystem;
Function	alpha-glucosidase activity; carbohydrate binding; hydrolase activity, hydrolyzing O-glycosyl compounds; maltose alpha-glucosidase activity;