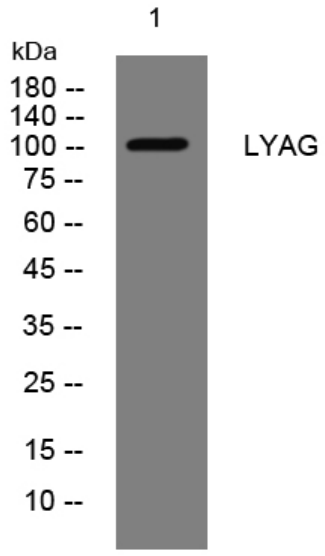
LYAG rabbit pAb

Catalog No	BYab-08106
Isotype	IgG
Reactivity	Human; Mouse;Rat
Applications	WB
Gene Name	GAA
Protein Name	LYAG
Immunogen	Synthesized peptide derived from human LYAG AA range: 432-482
Specificity	This antibody detects endogenous levels of LYAG at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.221% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	Lysosomal alpha-glucosidase (EC 3.2.1.20) (Acid maltase) (Aglucosidase alfa) [Cleaved into: 76 kDa lysosomal alpha-glucosidase; 70 kDa lysosomal alpha-glucosidase]
Observed Band	105kD
Cell Pathway	Lysosome . Lysosome membrane .
Tissue Specificity	Duodenum,Liver,Placenta,Plasma,Testis,Urine,
Function	catalytic activity:Hydrolysis of terminal, non-reducing (1->4)-linked alpha-D-glucose residues with release of alpha-D-glucose.,caution:The sequence shown here is derived from an Ensembl automatic analysis pipeline and should be considered as preliminary data.,disease:Defects in GAA are the cause of glycogen storage disease type 2 (GSD2) [MIM:232300]; also called acid alpha-glucosidase (GAA) deficiency or acid maltase deficiency (AMD). GSD2 is a metabolic disorder with a broad clinical spectrum. The severe infantile form, or Pompe disease, presents at birth with massive accumulation of glycogen in muscle, heart and liver. Cardiomyopathy and muscular hypotonia are the cardinal features of this form whose life expectancy is less than two years. The juvenile

	and adult forms present as limb-girdle muscular dystrophy beginning in the lower limbs. Final outcome depends on respiratory muscle fa
Background	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. [provided by RefSeq, Jan 2016],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



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