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Rabbit Anti-Galactosidase alpha antibody

SL7593R

Product Name:	Galactosidase alpha
Chinese Name:	α-半乳糖苷酶抗体
Alias:	Galactosidase alpha; Alpha D galactosidase A; Alpha D galactoside galactohydrolase; Melibiase; Alpha galactosidase A; GALA; GLA; GLA protein; AGAL_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	45kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Galactosidase alpha:101-200/429
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Galactosidase alpha is involved in the hydrolysis of terminal, non reducing alpha D galactose residues in alpha D galactosides, including galactose oligosaccharides, galactomannans and galactohydrolase. Defects in GLA are the cause of Fabry's disease (FD). FD is a rare X-linked sphingolipidosis disease where glycolipid accumulates in many tissues. Clinical recognition in males results from characteristic skin lesions

(angiokeratomas) over the lower trunk. Patients may show ocular deposits, febrile episodes, and burning pain in the extremities. Death results from renal failure, cardiac or cerebral complications of hypertension or other vascular disease. Heterozygous females may exhibit the disorder in an attenuated form, they are more likely to show corneal opacities.

Subunit:
Homodimer.

Subcellular Location:
Lysosome.

DISEASE:
Defects in GLA are the cause of Fabry disease (FD) [MIM:301500]. FD is a rare X-linked sphingolipidosis disease where glycolipid accumulates in many tissues. The disease consists of an inborn error of glycosphingolipid catabolism. FD patients show systemic accumulation of globotriaoslyceramide (Gb3) and related glycosphingolipids in the plasma and cellular lysosomes throughout the body. Clinical recognition in males results from characteristic skin lesions (angiokeratomas) over the lower trunk. Patients may show ocular deposits, febrile episodes, and burning pain in the extremities. Death results from renal failure, cardiac or cerebral complications of hypertension or other vascular disease. Heterozygous females may exhibit the disorder in an attenuated form, they are more likely to show corneal opacities.

Similarity:
Belongs to the glycosyl hydrolase 27 family.

SWISS:
P06280

Gene ID:
2717

Database links:
Entrez Gene: 2717Human

Entrez Gene: 11605Mouse

Entrez Gene: 363494Rat

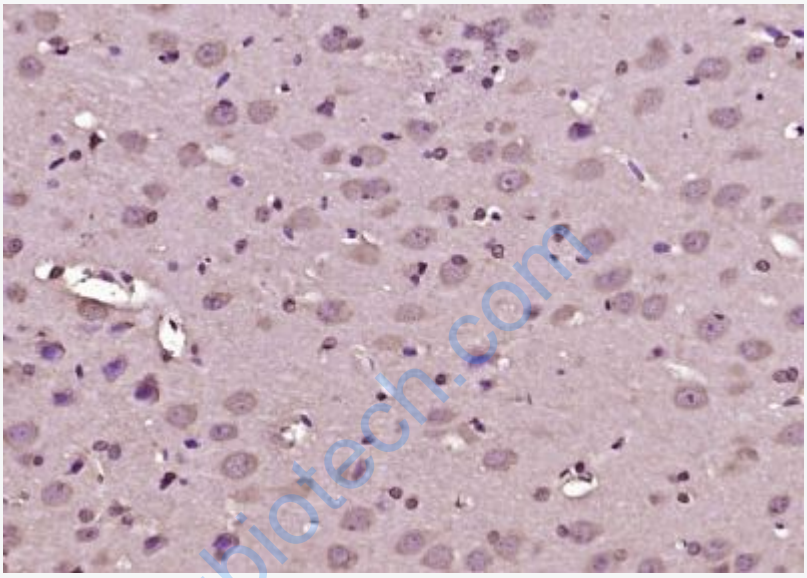
Omim: 300644Human

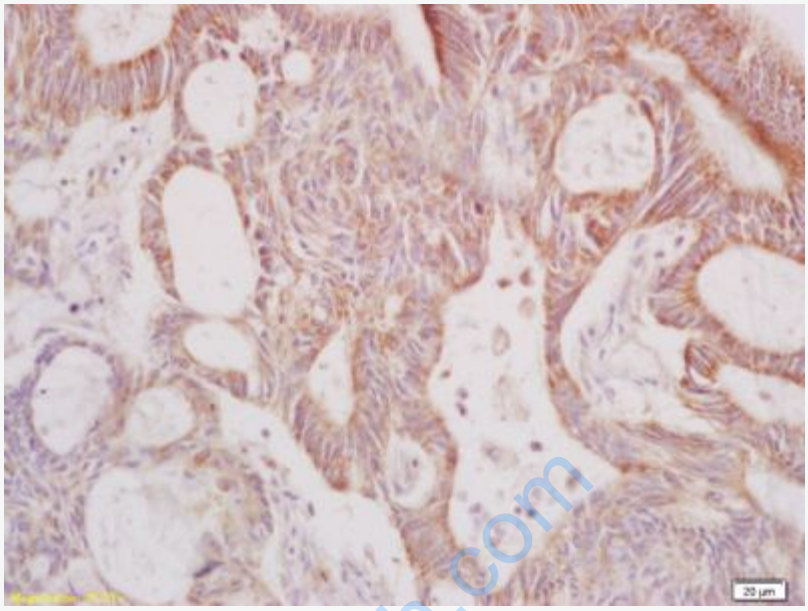
SwissProt: P06280Human

SwissProt: P51569Mouse

Unigene: 69089Human

Unigene: 1114Mouse

	Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Paraformaldehyde-fixed, paraffin embedded (rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Galactosidase alpha) Polyclonal Antibody, Unconjugated (SL7593R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.



Tissue/cell: human rectal carcinoma; 4% Paraformaldehyde-fixed and paraffin-embedded;

Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-Galactosidase alpha Polyclonal Antibody, Unconjugated(SL7593R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining