

Galactosidase alpha Rabbit pAb

Catalog # AP99076

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P06280
Other Accession	P06280
Reactivity	Rat, Human
Predicted	Pig, Mouse, Rabbit, Dog
Host	Rabbit
Clonality	Polyclonal
Calculated MW	48767
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Galactosidase alpha
Epitope Specificity	101-200/429
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the glycosyl hydrolase 27 family.
SUBUNIT	Homodimer.
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 globotriaosylceramide (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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	2717
Other Names	GALA; Ags; AGAL_HUMAN; GLA; Alpha-D-galactosidase A; Alpha-D-galactoside galactohydrolase; Galactosylgalactosylglucosylceramidase GLA; Melibiase; 3.2.1.22; AGAL_MOUSE; galactosidase alpha
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	GLA (HGNC:4296)
Function	Catalyzes the hydrolysis of glycosphingolipids and participates in their degradation in the lysosome.
Cellular Location	Lysosome.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.