abcam

Product datasheet

Recombinant Human Galactosidase alpha protein ab114549

1 图像

描述

产品名称	重组人Galactosidase alpha蛋白
表达系统	Wheat germ
Accession	P06280
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MQLRNPELHLGCALALRFLALVSWDIPGARALDNGLARTPTM GWLHWERF MCNLDCQEEPDSCISEKLFMEMAELMVSEGWKDAGYEYLCID DCWMAPQR DSEGRLQADPQRFPHGIRQLANYVHSKGLKLGIYADVGNKTC AGFPGSFG YYDIDAQTFADWGVDLLKFDGCYCDSLENLADGYKHMSLALN RTGRSIVY SCEWPLYMWPFQKPNYTEIRQYCNHWRNFADIDDSWKSIKSI LDWTSFNQ ERIVDVAGPGGWNDPDMLVIGNFGLSWNQQVTQMALWAIMAA PLFMSNDL RHISPQAKALLQDKDVIAINQDPLGKQGYQLRQGDNFEVWER PLSGLAWA VAMINRQEIGGPRSYTIAVASLGKGVACNPACFITQLLPVKR KLGFYEWT SRLRSHINPTGTVLLQLENTMQMSLKDLL
预测分子量	73 kDa including tags
氨基酸	1 to 429

技术指标

Our **Abpromise guarantee** covers the use of **ab114549** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	Western blot
	ELISA

	SDS-PAGE
形式	Liquid

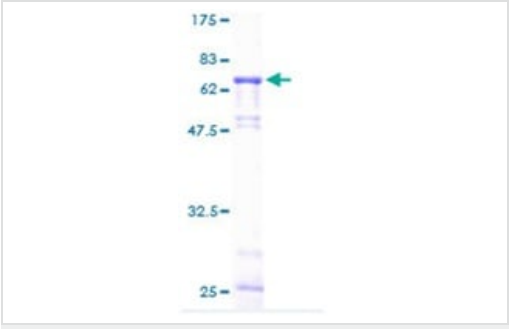
制备和贮存

稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

常规信息

疾病相关	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.
序列相似性	Belongs to the glycosyl hydrolase 27 family.
细胞定位	Lysosome.

图片



SDS-PAGE - Recombinant Human Galactosidase alpha protein (ab114549)

12.5% SDS-PAGE Stained with Coomassie Blue showing ab114549

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