

Product datasheet for TP724807

9620 Medical Center Drive, Ste 200 Rockville, MD 20850, US Phone: +1-888-267-4436 https://www.origene.com techsupport@origene.com

OriGene Technologies, Inc.

EU: info-de@origene.com CN: techsupport@origene.cn

GLB1 Human Recombinant Protein

Product data:

Product Type: Recombinant Proteins

Description: Recombinant Human beta-Galactosidase/GLB1(C-His)

Species: Human

Expression cDNA Clone

or AA Sequence:

Leu24-Val677

Tag: C-6His

Buffer: Lyophilized from a 0.2 um filtered solution of 20mM Tris-HCl, 150mM NaCl, pH 8.0

Note: Recombinant Human beta-Galactosidase is produced by our Mammalian expression system

and the target gene encoding Leu24-Val677 is expressed with a 6His tag at the C-terminus.

Stability: 12 months from date of despatch

Locus ID: 2720 **UniProt ID:** P16278

Summary: Î² Galactosidase is a lysosomal Î² Galactosidase that hydrolyzes the terminal Î² Galactose from

Ganglioside and Keratan sulfate. In lysosome, the mature \hat{l}^2 Galactosidase protein associates with Cathepsin A and Neuraminidase 1 to form the lysosomal multienzyme complex. An alternative splicing at the RNA level of \hat{l}^2 Galactosidase results a catalytically inactive \hat{l}^2 Galactosidase that plays an important role in vascular development. Defects of \hat{l}^2 -

galactosidase (GLB1) are the cause of diseases like GM1-gangliosidosis which is a lysosomal storage disease and Morquio Syndrome B that cause patients to have abnormal elastic fibers. More than 100 mutations have been identified for \hat{l}^2 Galactosidase, which result in different residual activities of the mutant enzymes and a spectrum of symptoms in the two related

diseases.

