

## 32-7321: Recombinant Human $\alpha$ -Galactosidase/GLA (C-6His)

**Gene :** GLA  
**Gene ID :** 2717  
**Uniprot ID :** P06280

### Description

Source: Human Cells.  
MW :46.39kD.

Recombinant Human  $\alpha$ -Galactosidase is produced by our Mammalian expression system and the target gene encoding Leu32-Leu429 is expressed with a 6His tag at the C-terminus.  $\alpha$ -Galactosidase A is a homodimeric glycoprotein that belongs to the glycosyl hydrolase 27 family. It is a lysosomal enzyme and used as a long-term enzyme replacement therapy in patients with a confirmed diagnosis of Fabry disease.  $\alpha$ -Galactosidase A can hydrolyze terminal  $\alpha$ -galactosyl moieties from glycolipids and glycoproteins and catalyze the hydrolysis of melibiose into galactose and glucose. Defects  $\alpha$ -Galactosidase A are the cause of Fabry disease (FD) which is a rare X-linked sphingolipidosis disease with 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. 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.

### Product Info

**Amount :** 10  $\mu$ g / 50  $\mu$ g  
**Content :** Supplied as a 0.2  $\mu$ m filtered solution of 20mM TrisHCl, 150mM NaCl, pH 8.0.  
**Storage condition :** Store at -20°C, stable for 6 months after receipt. Please minimize freeze-thaw cycles.  
**Amino Acid :** LDNGLARTPTMGWLHWERFMCNLDCQEEPDCISEKLFMEMAELMVSEGWKDAGYEYLCIDDCWMAPQRD  
SEGR LQADPQRFPHGIRQLANYVHSGKGLKGIYADVGNKTCAGFPGSFGYYDIDAQTFADWGVDLLKFDGICYC  
DSLENLADGYKHMSLALNRTGRSIVYSCEWPLYMWPFKPNYTEIRQYCNHWRNFADIDDSWKSILSDWTS  
FNQERIVDVAGPGGWNDPMLVIGNFGLSWNQVQTMALWAIMAAPLFMSNDLRHISPOAKALLQDKDVIAI  
NQDPLGKQGYQLRQGDNFEVWERPLSGLAWAVAMINRQEIGGPRSYTIAVASLGKGVACNPACFITQLLPVKR  
KLGFEWTSRLRSHINPTGTVLLQLENTMQMSLKDLLVDHHHHHH

### Application Note

**Endotoxin :** Less than 0.1 ng/ $\mu$ g (1 IEU/ $\mu$ g) as determined by LAL test.