

Native α -1,2-Fucosidase solution

Cat. No. NATE-0259

Lot. No. (See product label)

Introduction

Description

Tissue alpha-L-fucosidase is an enzyme that in humans is encoded by the FUCA1 gene. Alpha-Fucosidase is an enzyme that breaks down fucose. Fucosidosis is an autosomal recessive lysosomal storage disease caused by defective alpha-L-fucosidase with accumulation of fucose in the tissues. Different phenotypes include clinical features such as neurologic deterioration, growth retardation, visceromegaly, and seizures in a severe early form; coarse facial features, angiokeratoma corporis diffusum, spasticity and delayed psychomotor development in a longer surviving form; and an unusual spondylometaphyseal dysplasia in yet another form.

Synonyms

α -1,2-Fucosidase; alpha-L-fucosidase; Alpha-Fucosidase; FUCA1; FUCA; 9037-65-4

Product Information

Form

buffered aqueous solution, Solution in 20 mM Tris-HCl, pH 7.5, 25 mM NaCl

EC Number

EC 3.2.1.51

CAS No.

9037-65-4

Activity

> 0.4 units/mg protein

Pathway

Lysosome, organism-specific biosystem; Other glycan degradation, organism-specific biosystem

Function

alpha-L-fucosidase activity; carbohydrate binding

Unit Definition

One unit will release 1.0 μ mole of fucose from 2'-fucosyllactose per min at pH 5.0 at 37°C

Usage and Packaging

Package

vial of 0.004 unit

Storage and Shipping Information

Storage

2-8°C