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Recombinant Human ATL1/SPG3A/Atlastin-1 Protein (GST Tag)

Catalog Number: PKSH031549

Note: Centrifuge before opening to ensure complete recovery of vial contents.

Description

Species Human

Source Baculovirus-Insect Cells-derived Human ATL1/SPG3A/Atlastin-1 protein Met 1-Thr

447, with an N-terminal GST

Calculated MW 77.0 kDa Observed MW 66 kDa Accession NP 056999.2

Not validated for activity **Bio-activity**

Properties

> 80 % as determined by reducing SDS-PAGE. **Purity**

Endotoxin < 1.0 EU per µg of the protein as determined by the LAL method.

Storage Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80

°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of

reconstituted samples are stable at < -20°C for 3 months.

This product is provided as lyophilized powder which is shipped with ice packs. Shipping Lyophilized from sterile 50mM Tris, 100mM NaCl, 0.5mM PMSF, 0.5mM EDTA, **Formulation**

0.5mM GSH, pH 8.0

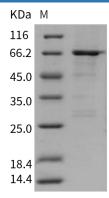
Normally 5% - 8% trehalose, mannitol and 0.01% Tween 80 are added as protectants

before lyophilization.

Please refer to the specific buffer information in the printed manual.

Reconstitution Please refer to the printed manual for detailed information.

Data



> 80 % as determined by reducing SDS-PAGE.

Background

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Atlastin-1, also known as Spastic paraplegia 3 protein A, Guanine nucleotide-binding protein 3, GTP-binding protein 3, GBP3, ATL1 and SPG3A, is a multi-pass membrane protein which belongs to the GBP family and atlastin subfamily. ATL1 / SPG3A is expressed predominantly in the adult and fetal central nervous system. Expression of ATL1 / SPG3A in adult brain is at least 50-fold higher than in other tissues. ATL1 / SPG3A is detected predominantly in pyramidal neurons in the cerebral cortex and the hippocampus of the brain. ATL1 / SPG3A is also expressed in upper and lower motor neurons (at protein level). A distinguishing feature of ATL1 / SPG3A is its frequent early onset, raising the possibility that developmental abnormalities may be involved in its pathogenesis. Missense SPG3A mutant atlastin-1 proteins have impaired GTPase activity and may act in a dominant-negative, loss-of-function manner by forming mixed oligomers with wild-type atlastin-1. Defects in ATL1 / SPG3A are the cause of spastic paraplegia autosomal dominant type 3 (SPG3), also known as Strumpell-Lorrain syndrome. Spastic paraplegia is a degenerative spinal cord disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs.

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