

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

Catalog Number: 10523-H09B



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Biological Solution Specialist

General Information

Gene Name Synonym:

AD-FSP; atlastin1; FSP1; GBP3; HSN1D; SPG3; SPG3A; 4930435M24Rik; Adfsp; Fsp1; Spg3; Spg3a

Protein Construction:

A DNA sequence encoding the human SPG3A (NP_056999.2) (Met 1-Thr 447) was fused with the GST tag at the N-terminus.

Source: Human

Expression Host: Baculovirus-Insect Cells

QC Testing

Purity: > 80 % as determined by SDS-PAGE

Endotoxin:

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

Stability:

Samples are stable for up to twelve months from date of receipt at -70 °C

Predicted N terminal: Met

Molecular Mass:

The recombinant human SPG3A/GST chimera consists of 671 amino acids and has a calculated molecular mass of 77 kDa. It migrates as an approximately 66 kDa band in SDS-PAGE under reducing conditions.

Formulation:

Lyophilized from sterile 50mM Tris, 100mM NaCl, 0.5mM PMSF, 0.5mM EDTA, 0.5mM GSH, pH 8.0

Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Specific concentrations are included in the hardcopy of COA. Please contact us for any concerns or special requirements.

Usage Guide

Storage:

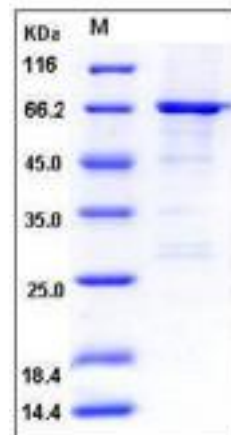
Store it under sterile conditions at -20°C to -80°C upon receiving. Recommend to aliquot the protein into smaller quantities for optimal storage.

Avoid repeated freeze-thaw cycles.

Reconstitution:

Detailed reconstitution instructions are sent along with the products.

SDS-PAGE:



Protein Description

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.

References

1. Zhao X., *et al.*, (2001), Mutations in a newly identified GTPase gene cause autosomal dominant hereditary spastic paraplegia. *Nat. Genet.* 29:326-331.
2. Luan Z., *et al.*, (2002), A novel GTP-binding protein hGBP3 interacts with NIK/HGK. *FEBS Lett.* 530:233-238.
3. Ota T., *et al.*, (2004), Complete sequencing and characterization of 21,243 full-length human cDNAs. *Nat. Genet.* 36:40-45.

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