

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



Catalog Number:PKSH031549

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

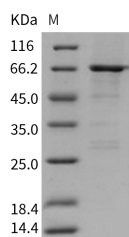
Description

| | |
|------------------------------------|---|
| Synonyms | AD-FSP;atlastin1;FSP1;GBP3;HSN1D;SPG3;SPG3A |
| Species | Human |
| Expression Host | Baculovirus-Insect Cells |
| Sequence | Met 1-Thr 447 |
| Accession | NP_056999.2 |
| Calculated Molecular Weight | 77.0 kDa |
| Observed molecular weight | 66 kDa |
| Tag | N-GST |

Properties

| | |
|-----------------------|--|
| Purity | > 80 % as determined by reducing SDS-PAGE. |
| 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. |
| Shipping | This product is provided as lyophilized powder which is shipped with ice packs. |
| 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. Please refer to the specific buffer information in t |
| Reconstitution | Please refer to the printed manual for detailed information. |

Data



> 80 % as determined by reducing SDS-PAGE.

Background

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

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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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