

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

Catalog No. PKSH031549

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

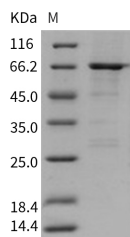
Description

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
Bioactivity	Not validated for activity

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% 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

Atlastin-1, also known as Spastic paraplegia 3 protein A, Guanine nucleotide-binding protein 3, GTP-binding protein 3,

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GBP3, ATL1 and SPG3A, is a multi-pass membrane protein which belongs to theGBP 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.