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

**Cat.NO.: TP06620**

3th Edition

**Synonyms:**AD-FSP;atlastin1;FSP1;GBP3;HSN1D;SPG3;SPG3A

**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.

**Form:**PBS

**Molecular Weight:**77 kDa

**Sequences:**Met 1-Thr 447

**Purity:**> 95% by HPLC

**Concentration:**

**Endotoxin Level:**<1.0 EU per 1 ug of protein (determined by LAL method)

**Storage:**Can be stored at +4°C short term (1-2 weeks). For long term storage, aliquot and store at -20°C or -70°C. Avoid repeated freezing and thawing cycles.