

Recombinant Protein Technical Manual Recombinant Human UBE1/UBA1 Protein (His & GST Tag) RPES4270

Product Data:

Product SKU: RPES4270 **Size:** 20μg

Species: Human Expression host: Baculovirus-Insect Cells

Uniprot: NP 003325.2

Protein Information:

Molecular Mass: 146 kDa

AP Molecular Mass: 130 kDa

Tag: N-His & GST

Bio-activity:

Purity: > 96 % as determined by reducing SDS-PAGE.

Endotoxin: $< 1.0 \text{ EU per } \mu\text{g}$ as determined by the LAL method.

Storage: 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, pH 7.4, 10% gly, 0.5mM GSH

Reconstitution: Please refer to the printed manual for detailed information.

Application:

Synonyms: A1S9;A1S9T;A1ST;AMCX1;CFAP124;CTD-

2522E6.1;GXP1;POC20;SMAX2;UBA1A;UBE1;UBE1X

Immunogen Information:

Sequence: Ser 2-Arg 1058

Background:

UBE1, also known as UBA1, belongs to the ubiquitin-activating E1 family. UBE1 gene complements an X-linked mouse temperature-sensitive defect in DNA synthesis, and thus may function in DNA repair. It is part of a gene cluster on chromosome Xp11.23. UBE1 catalyzes the first step in ubiquitin conjugation to mark cellular proteins for degradation. It also catalyzes the first step in ubiquitin conjugation to mark cellular proteins for degradation by first adenylating its C-terminal glycine residue with ATP, and thereafter linking this residue to the side chain of a cysteine residue in E1, yielding an ubiquitin-E1 thioester and free AMP. Defects in UBA1 can cause spinal muscular atrophy X-linked type 2 (SMAX2), also known as X-linked lethal infantile spinal muscular atrophy, distal X-linked arthrogryposis multiplex congenita or X-linked arthrogryposis type 1 (AMCX1). Spinal muscular atrophy refers to a group of neuromuscular disorders characterized by degeneration of the anterior horn cells of the spinal cord, leading to symmetrical muscle weakness and atrophy. SMAX2 is a lethal infantile form presenting with hypotonia, areflexia, and multiple congenital contractures.