

## Recombinant Human UBE1/UBA1 Protein (His & GST Tag)

Catalog No. PKSH030942

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

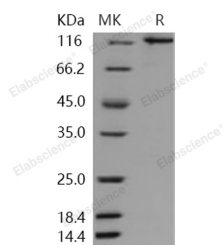
### Description

<b>Synonyms</b>	A1S9;A1S9T;A1ST;AMCX1;CFAP124;CTD-2522E6.1;GXP1;POC20;SMAX2;UBA1A;UBE1;UBE1X
<b>Species</b>	Human
<b>Expression Host</b>	Baculovirus-Insect Cells
<b>Sequence</b>	Ser 2-Arg 1058
<b>Accession</b>	NP_003325.2
<b>Calculated Molecular Weight</b>	146 kDa
<b>Observed molecular weight</b>	130 kDa
<b>Tag</b>	N-His-GST
<b>Bioactivity</b>	Not validated for activity

### Properties

<b>Purity</b>	> 96 % as determined by reducing SDS-PAGE.
<b>Endotoxin</b>	< 1.0 EU per µg of the protein as determined by the LAL method.
<b>Storage</b>	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.
<b>Shipping</b>	This product is provided as lyophilized powder which is shipped with ice packs.
<b>Formulation</b>	Lyophilized from sterile 50mM Tris, 100mM NaCl, pH 7.4, 10% glycerol, 0.5mM GSH 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.
<b>Reconstitution</b>	Please refer to the printed manual for detailed information.

### Data



> 96 % as determined by reducing SDS-PAGE.

### Background

#### For Research Use Only

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.