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**Recombinant Human UBE1 / UBA1 Protein (His & GST tag)****Cat.NO.: TP06299**

3th Edition

**Synonyms:**A1S9;A1S9T;A1ST;AMCX1;CFAP124;CTD-2522E6.1;GXP1;POC20;SMAX2;UBA1A;UBE1;UBE1X

**Description:**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 arthrogyrosis multiplex congenita or X-linked arthrogyrosis 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.

**Form:**PBS**Molecular Weight:**146 kDa**Sequences:**Ser 2-Arg 1058**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.