

Cat. No:	ABN19503
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from the N-terminal region of human UBA1. AA range:91-140
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,ELISA 1:10000-1:20000
Molecular Weight:	118kDa
Purification:	Affinity purification
Synonyms:	UBA1; A1S9T; UBE1; Ubiquitin-like modifier-activating enzyme 1; Protein A1S9; Ubiquitin-activating enzyme E1

Background:	<p>The protein encoded by this gene catalyzes the first step in ubiquitin conjugation to mark cellular proteins for degradation. This 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. Alternatively spliced transcript variants that encode the same protein have been described. [provided by RefSeq, Jul 2008],disease:Defects in UBA1 are the cause of spinal muscular atrophy X-linked type 2 (SMAX2) [MIM:301830]; 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.,function:Activates ubiquitin 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.,miscellaneous:There are two active sites within the E1 molecule, allowing it to accommodate two ubiquitin moieties at a time, with a new ubiquitin forming an adenylate intermediate as the previous one is transferred to the thiol site.,pathway:Protein modification; protein ubiquitination.,similarity:Belongs to the ubiquitin-activating E1 family.,subunit:Monomer (By similarity). Interacts with GAN (via BTB domain).</p>
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

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