



## Anti-UBA1 Antibody

**Alternative Names:** A1S9, A1ST, GXP1, UBE1, A1S9T, AMCX1, POC20, SMAX2, UBA1A, UBE1X, CFAP124, Ubiquitin Like Modifier Activating Enzyme, Ubiquitin-Activating Enzyme E1, A1S9T And BN75 Temperature Sensitivity Complementing, Ubiquitin-Activating Enzyme E1 Homolog, Testicular

**Catalogue Number:** AB19-10122-100ug

**Size:** 100 µg

## Background Information

Ubiquitin-like modifier-activating enzyme 1 (UBA1) is a multidomain enzyme and regulator of the ubiquitin proteasome system (UPS). It effectively marks proteins for degradation by the proteasome, acting as a gatekeeper of the Ub conjugation cascade by activating ubiquitin in a two-step process involving adenylation and thioester bond formation followed by transfer of ubiquitin to Cdc34 (an E2 ubiquitin-conjugating enzyme). Mutations in the ubiquitin-like modifier activating enzyme 1 gene (UBA1), leading to reduced UBA1 expression, cause a rare form of Spinal Muscular Atrophy (SMA) known as X-linked SMA (XL-SMA). Aminoacyl-tRNA synthetases, including GARS (Glycyl-tRNA synthetase), have been identified as downstream targets of UBA1. Dysregulation of UBA1/GARS pathways in spinal muscular atrophy results in a similar phenotype to GARS-dependent defects (such as Charcot-Marie-Tooth disease), perhaps indicating significant molecular and phenotypic overlap between spinal muscular atrophy and Charcot-Marie-Tooth disease.

## Product Information

<b>Antibody Type:</b>	Polyclonal	<b>Host:</b>	Rabbit
<b>Isotype:</b>	IgG	<b>Species Reactivity:</b>	Human, Mouse, Rat
<b>Immunogen:</b>	A synthetic peptide from human UBA1		
<b>Format:</b>	100 µg in 100 µl PBS with 0.03% Proclin300, 50% glycerol, pH7.3.		
<b>Storage Conditions:</b>	Store at -20°C. Avoid freeze / thaw cycles.		
<b>Applications:</b>	WB IHC WB 1:500-2000. IHC 1:30-150.		

## Additional Information

<b>Subcellular location:</b>	Nucleus, Mitochondrion	<b>MW:</b>	118 kDa (Intended as a general guide and does not allow for all isoforms and species variations)
<b>Gene ID</b>	7317	<b>Uniprot ID:</b>	P22314