

## Recombinant Human UPB1, His-tagged

Cat. No. UPB1-49H Lot. No. (See product label)

### SPECIFICATION

**Product Overview** Recombinant Human  $\beta$ -Ureidopropionase/UPB1 is produced by our E. coli expression system. The target protein is expressed with sequence (Met1-Glu384) of Human UPB1 fused with a His tag at the C-terminus.

**Species** Human

**Source** E.coli

**ProteinLength** 1-384 a.a.

**Description**  $\beta$ -Ureidopropionase is a cytoplasmic protein which belongs to the CN hydrolase family of BUP subfamily.  $\beta$ -Ureidopropionase binds one zinc ion per subunit, catalyzes the last step in the pyrimidine degradation pathway.  $\beta$ -Ureidopropionase can convert N-carbamyl-beta-aminoisobutyric acid and N-carbamyl-beta-alanine to beta-aminoisobutyric acid and beta-alanine, ammonia and carbon dioxide, respectively. The pyrimidine bases uracil and thymine are degraded via the consecutive action of dihydropyrimidine dehydrogenase (DHPDH), dihydropyrimidinase (DHP) and beta-ureidopropionase (UP) to beta-alanine and beta aminoisobutyric acid, respectively. Defects in  $\beta$ -Ureidopropionase are the cause of  $\beta$ -Ureidopropionase deficiency that is characterized by muscular hypotonia, dystonic movements, scoliosis, microcephaly and severe developmental delay.

**Form** Supplied as a 0.2  $\mu$ M filtered solution of 20mM PB, 150mM NaCl, pH 7.4

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**AA Sequence**

MAGAEWKSLEECLEKHLPLPDLQEVRVLYGKELRKLDLPREAFEASREDFELQG  
 YAFEAAEEQ LRRPRIVHVGLVQNRIPANAPVAEQVSALHRRIKAIVEVAAMCGVNI  
 ICFQEAWTMPFAFCTR EKLPWTEFAESAEDGPTTRFCQKLAKNHDMVVVSPILERD  
 SEHGDVLWNTAVVISNSGAVLGKTR KNHIPRVGDFNESTYYMEGNLGHPVFQTQF  
 GRIAVNICYGRHHPLNWL MYSINGAEIIFNPSATI GALSESLWPIEARNAAIANHCFTC  
 AINRVGTEHFPNEFTSGDGKKAHQDFGYFYGSSYVAAPDSS RTPGLSRSRDGLLV  
 AKLDLNLCCQVNDVWNFKMTGRYEMYARELAEAVKSNYSPTIVKEVEHHHH HH

**Endotoxin** Less than 0.1 ng/μg (1 IEU/μg).

**Purity** Greater than 95% as determined by reducing SDS-PAGE.

**Storage** Store at Please minimize freeze-thaw cycles.

## GENE INFORMATION

**Gene Name** [UPB1 ureidopropionase, beta \[ Homo sapiens \]](#)

**Official Symbol** UPB1

**Synonyms** UPB1; ureidopropionase, beta; beta-ureidopropionase; BUP1; BUP-1; beta-alanine synthase; n-carbamoyl-beta-alanine amidohydrolase;

**Gene ID** [51733](#)

**mRNA Refseq** [NM\\_016327](#)

**Protein Refseq** [NP\\_057411](#)

**MIM** [606673](#)

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<b>UniProt ID</b>	Q9UBR1
<b>Chromosome Location</b>	22q11.2
<b>Pathway</b>	Drug metabolism - other enzymes, organism-specific biosystem; Drug metabolism - other enzymes, conserved biosystem; Fluoropyrimidine Activity, organism-specific biosystem; Metabolic pathways, organism-specific biosystem; Metabolism, organism-specific biosystem; Metabolism of nucleotides, organism-specific biosystem; Pantothenate and CoA biosynthesis, organism-specific biosystem;
<b>Function</b>	beta-ureidopropionase activity; hydrolase activity, acting on carbon-nitrogen (but not peptide) bonds; metal ion binding;

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