## Recombinant Human BUP1 Protein (His Tag)

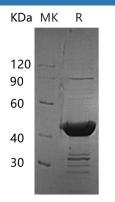
Note: Centrifuge before opening to ensure complete recovery of vial contents.

Catalog Number: PKSH033271



Description **Species** Human 44.2 kDa Mol Mass Accession O9UBR1 Not validated for activity **Bio-activity Properties** > 95 % as determined by reducing SDS-PAGE. Purity Endotoxin < 1.0 EU per µg of the protein as determined by the LAL method. Storage Store at  $< -20^{\circ}$ C, stable for 6 months. Please minimize freeze-thaw cycles. This product is provided as liquid. It is shipped at frozen temperature with blue ice/gel Shipping packs. Upon receipt, store it immediately at  $< -20^{\circ}$ C. Formulation Supplied as a 0.2 µm filtered solution of PBS, pH7.4. Reconstitution Not Applicable

## Data



> 95 % as determined by reducing SDS-PAGE.

## Background

 $\beta$ -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 betaaminoisobutyric 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.

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