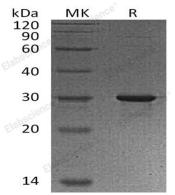
Recombinant Human GAMT Protein (His Tag)

Catalog Number: PKSH032515

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

Description	
Species	Human
Source	E.coli-derived Human GAMT protein Met 1-Gly236, with an N-terminal His & C-
	terminal His
Calculated MW	29.5 kDa
Observed MW	27-32 kDa
Accession	Q14353
Bio-activity	Not validated for activity
Properties	
Purity	> 90 % as determined by reducing SDS-PAGE.
Concentration	Subject to label value.
Endotoxin	< 1.0 EU per µg of the protein as determined by the LAL method.
Storage	Store at $<$ -20°C, stable for 6 months. Please minimize freeze-thaw cycles.
Shipping	This product is provided as liquid. It is shipped at frozen temperature with blue ice/gel
	packs. Upon receipt, store it immediately at $< -20^{\circ}$ C.
Formulation	Supplied as a 0.2 µm filtered solution of 20mM Tris-HCl, 1mM DTT, pH 8.0.

Data



> 90 % as determined by reducing SDS-PAGE.

Background

GAMT is a methyltransferase which belongs to the class I-like SAM-binding methyltransferase superfamily. It contains one RMT2 (arginine N-methyltransferase 2-like) domain and is expressed in liver. GAMT converts guanidoacetate to creatine, using S-adenosylmethionine as the methyl donor. Defects in GAMT are the cause of guanidinoacetate methyltransferase deficiency, which is an autosomal recessive disorder characterized by developmental delay/regressio n, mental retardation, severe disturbance of expressive and cognitive speech, intractable seizures and movement disturbances, severe depletion of creatine/phosphocreatine in the brain, and accumulation of guanidinoacetic acid in brain and body fluids.