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Recombinant Human MOGS Protein, N-His

Catalog Number:	bs-105596P
Species:	Human
AA Seq:	360-485/837
Predicted MW:	16.14 kDa
Tags:	N-His
Activity:	Not tested
Purity:	>90% as determined by SDS-PAGE.
Purification:	AC
Form:	Lyophilized
Storage:	Lyophilized from a solution in PBS pH 7.4, 0.02% NLS, 1mM EDTA, 4% Trehalose, 1% Mannitol. Use a manual defrost freezer and avoid repeated freeze thaw cycles. Store at 2 to 8°C for frequent use. Store at -20 to -80°C for twelve months from the date of receipt.
Background:	Glycosylation of asparagine residues in Asn-X-Ser/Thr motifs in proteins commonly occur in the lumen of the endoplasmic reticulum (ER). Glucosidase I catalyzes the first step in the N-linked oligosaccharide processing pathway. It specifically removes the distal alpha 1,2-linked glucose residue from the Glc3-Man9-GlcNAc2 oligosaccharide precursor. Glucosidase I contains a short cytosolic tail, a single pass transmembrane domain and a large C-terminal catalytic domain located on the luminal side of the ER. Mutations in the gene encoding Glucosidase I result in the congenital disorder glycosylation (CDG-IIb), which is characterized by generalized hypotonia, dysmorphic features, hepatomegaly, hypoventilation, feeding problems, seizures and death. Two point mutations in the Glucosidase I gene have been identified and result in amino acid substitutions, namely Arg486Thr and Phe652Leu, that affect polypeptide folding and active site formation.