## Recombinant Human OSTM1 Protein (His Tag)

## Catalog Number: PKSH031358

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

Description			
Species	Human		
Source	HEK293 Cells-derived Human OSTM1 protein Met 1-Pro 284, with an C-terminal His		
Calculated MW	29.7 kDa		
Observed MW	40-50 kDa		
Accession	NP_054747.2		
Bio-activity	Not validated for activity		
Properties			
Purity	> 97 % as determined by reducing SDS-PAGE.		
Endotoxin	< 1.0 EU per µg of the protein as determined by the LAL method.		
Storage	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80		
	°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of		
	reconstituted samples are stable at $< -20^{\circ}C$ for 3 months.		
Shipping	This product is provided as lyophilized powder which is shipped with ice packs.		
Formulation	Lyophilized from sterile PBS, pH 7.4		
	Normally 5% - 8% trehalose, mannitol and 0.01% Tween 80 are added as protectants		
	before lyophilization.		
	Please refer to the specific buffer information in the printed manual.		
Reconstitution	Please refer to the printed manual for detailed information.		



KDa	MK	R
116 66.2	=	_
45.0 35.0	-	
25.0	-	
18.4 14.4	=	

> 97 % as determined by reducing SDS-PAGE.

## Background

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Osteopetrosis-associated transmembrane protein 1 (OSTM1) is a Single-pass type I membrane protein. It is expressed in many hematopoietic cells of the myeloid and lymphoid B- and T-lineages. The analysis of OSTM1 association with CLCN7 demonstrated that OSTM1 requires CLCN7 to localize to lysosomes, whereas the formation of a CLCN7-OSTM1 complex is required to stabilize CLCN7. The researches found that OSTM1 plays a major role in myelopoiesis and lymphopoiesis and provided evidence of a crosstalk mechanism between hematopoietic cells for osteoclast activation. Thus, OSTM1 has a important role in osteoclast function and activation. The loss of function of OSTM1 results in deregulation of multiple hematopoietic lineages in addition to osteoclast lineage, OSTM1-defect patients display the most severe recessive osteopetrotic phenotype and die at early ages. Furthermore, it is suggested that OSTM1 has a primary role in neural development not related to lysosomal dysfunction. The canonical Wnt/beta-catenin signaling pathway may be a molecular basis for OSTM1 mutations and severe autosomal recessive osteopetrosis (ARO).