## Recombinant Human FLNC protein (His Tag)

## Catalog Number: PDEH100853

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

Description		
Species	Human	
Source	E.coli-derived Human FLNC protein Thr2519-Pro2725, with an N-terminal His	
Calculated MW	22.7 kDa	
Observed MW	28 kDa	
Accession	Q14315	
Bio-activity	Not validated for activity	
Properties		
Purity	> 95% as determined by reducing SDS-PAGE.	
Endotoxin	< 10 EU/mg 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 a 0.2 $\mu m$ filtered solution in PBS with 5% Trehalose and 5%	
	Mannitol.	
Reconstitution	It is recommended that sterile water be added to the vial to prepare a stock solution of	
	0.5 mg/mL. Concentration is measured by UV-Vis.	



KDa	М	R
80 60	-	
40		
30	-	_
20		
12	-	

> 95 % as determined by reducing SDS-PAGE.

## Background

FLNC is a muscle-specific filamin, which plays a central role in muscle cells, probably by functioning as a large actincross-linking protein. May be involved in reorganizing the actin cytoskeleton in response to signaling events, and may also display structural functions at the Z-disks in muscle cells. Defects in FLNC are the cause of autosomal dominant filaminopathy. Myofibrillar myopathy (MFM) is a neuromuscular disorder, usually with an adult onset, characterized by focal myofibrillar destruction and pathological cytoplasmic protein aggregations. Autosomal dominant filaminopathy is a form of MFM characterized by morphological features of MFM and clinical features of a limb-girdle myopathy. A heterozygous nonsense mutation which segregates with the disease, has been identified in the FLNC gene.

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