DST / Dystonin (DST) Human Recombinant Protein

PRODUCT OVERVIEW

Product Name	DST / Dystonin, Recombinant Protein
Full Product Name	Human DST / Dystonin Recombinant Protein
Product Gene Name	DST recombinant protein
Product Synonym Names	Dystonin; 230 kDa bullous pemphigoid antigen; 230/240 kDa bullous pemphigoid antigen; Bullous pemphigoid antigen 1; BPA; Dystonia musculorum protein; Hemidesmosomal plaque protein; BP230; BP240; BPAG1; DMH; DT; KIAA0728
Host	E Coli
Form/Format	Liquid; 50mM NaH2PO4, 500mM NaCl Buffer with 500mM Imidazole, 10% Glycerol (PH8.0)
Purity/Purification	>95% by SDS-PAGE
Concentration	Reconstitution Dependent (lot specific)
Preparation and Storage	Store at -20 degree C. Avoid repeated freezing and thawing.
OMIM	phenotype 615425
3D Structure	ModBase 3D Structure for Q03001
Species	Human
Tag	His
Subunit	Homodimer. Isoform 1 interacts (via N-terminus) with PLEC (via N-terminus). Interacts with the neuronal intermediate filament protein, PRPH. Interacts with DES. Interacts with SYNE3 (By similarity). Isoform 1 and isoform 6 can homodimerize (via N-terminus). Isoform 1 interacts (via N-terminus) with ACTN2. Isoform 1 interacts (via N-terminus) with PLEC (via N-terminus). Isoform 3 interacts (via N-terminus) with COL17A1 (via cytoplasmic region). Isoform 3 interacts (via N-terminus) with ITGB4 isoform beta-4a (via cytoplasmic region). Isoform 3 interacts (via N-terminus) with ERBIN (via C-terminus). Isoform 3 associates (via C-terminal) with KRT5-KRT14 (via rod region) intermediate filaments of keratins. Interacts with MAPRE1; probably required for targeting to the growing microtubule plus ends. Interacts with TMIGD2. Isoform 9 interacts with TMEM108.
Entry Function	Isoform 7: regulates the organization and stability of the microtubule network of sensory neurons to allow axonal transport.
Applications Tested/Suitable for DST recombinant protein	ELISA (EIA), Western Blot (WB), Antigen Protein (AP), Protein Array
Related Product Information for DST recombinant protein	MBS2902751 is a recombinant protein targeting DST. Also known as Human DST / Dystonin Recombinant Protein. Host: E Coli.
Other Notes	Small volumes of DST recombinant protein vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for DST. It may not necessarily be applicable to this product.

NCBI GI #	294862529
NCBI GeneID	667
NCBI Accession #	Q03001.4

UniProt Primary Accession #	Q03001
UniProt Related Accession #	Q03001
NCBI Official Full Name	Dystonin
NCBI Official Synonym Full Names	dystonin
NCBI Official Symbol	DST
NCBI Official Synonym Symbols	DT; BPA; DMH; BP240; BPAG1; EBSB2; HSAN6; MACF2; CATX15; CATX-15; D6S1101
UniProt Protein Name	Dystonin
UniProt Synonym Protein Names	230 kDa bullous pemphigoid antigen; 230/240 kDa bullous pemphigoid antigen; Bullous pemphigoid antigen 1; BPA; Bullous pemphigoid antigen; Dystonia musculorum protein; Hemidesmosomal plaque protein
Protein Family	Dystonin
UniProt Gene Name	DST
UniProt Synonym Gene Names	BP230; BP240; BPAG1; DMH; DT; KIAA0728; BPA; Bullous pemphigoid antigen
UniProt Entry Name	DYST_HUMAN
NCBI Protein Information	dystonin
NCBI Summary for DST	This gene encodes a member of the plakin protein family of adhesion junction plaque proteins. Multiple alternatively spliced transcript variants encoding distinct isoforms have been found for this gene, but the full-length nature of some variants has not been defined. It has been reported that some isoforms are expressed in neural and muscle tissue, anchoring neural intermediate filaments to the actin cytoskeleton, and some isoforms are expressed in epithelial tissue, anchoring keratin-containing intermediate filaments to hemidesmosomes. Consistent with the expression, mice defective for this gene show skin blistering and neurodegeneration. [provided by RefSeq, Mar 2010]
UniProt Comments for DST	BPAG1 iso3: a protein of the plakin family. Plakins are multi-domain proteins that have been shown to interact with microtubules, actin filaments and intermediate filaments, as well as proteins found in cellular junctions. Ten alternatively spliced human isoforms have been reported. BPAG1 has neuronal and muscle isoforms that consist of actin-binding and microtubule-binding domains at either end separated by a plakin domain and many spectrin repeats. Bpag1 isoforms with different N-termini may have differing roles. The plakin domain contains an apparent nuclear localization signal. Protein type: Microtubule-binding; Cytoskeletal; Motility/polarity/chemotaxis; Actin-binding Chromosomal Location of Human Ortholog: 6p12.1 Cellular Component: endoplasmic reticulum membrane; intermediate filament cytoskeleton; focal adhesion; cytoplasmic membrane-bound vesicle; leading edge; integral to membrane; intermediate filament; nuclear envelope; cell cortex; Z disc; cytosol; actin cytoskeleton; microtubule cytoskeleton; hemidesmosome; axon; cytoplasm; basal plasma membrane; basement membrane; nucleus Molecular Function: protein C-terminus binding; integrin binding; microtubule plus-end binding; protein binding; protein homodimerization activity; calcium ion binding; actin binding Biological Process: integrin-mediated signaling pathway; hemidesmosome assembly; extracellular matrix organization and biogenesis; cell motility involved in cell locomotion; response to wounding; intermediate filament cytoskeleton organization and biogenesis; cytoskeleton organization and biogenesis; microtubule cytoskeleton organization and biogenesis; cell cycle arrest; cell adhesion; maintenance of cell polarity
Research Articles on DST	**1.** A report on 2 novel heterozygous mutations in the dystonin (DST) gene from a family with hereditary sensory and autonomic neuropathy type VI. Induced-pluripotent stem cells findings suggest that the dystonin defect might alter proper development of the peripheral nerves

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