

Plectin Rabbit pAb

db2082

Package : 20µL 50µL 100µL

Product Name : Plectin Rabbit pAb**Cat.No.:** db2082**Synonyms** : HD1; PCN; EBS1; EBSO; PLTN; EBSMD; EBSND; EBSOG; EBSPA; PLEC1; LGMD2Q; PLEC1b**Application** : WB, IHC, IP**Reactivity** : Human, Mouse, Rat**Host species** : Rabbit**Background**

Plectin is a prominent member of an important family of structurally and in part functionally related proteins, termed plakins or cytolinkers, that are capable of interlinking different elements of the cytoskeleton. Plakins, with their multi-domain structure and enormous size, not only play crucial roles in maintaining cell and tissue integrity and orchestrating dynamic changes in cytoarchitecture and cell shape, but also serve as scaffolding platforms for the assembly, positioning, and regulation of signaling complexes (reviewed in PMID: 9701547, 11854008, and 17499243). Plectin is expressed as several protein isoforms in a wide range of cell types and tissues from a single gene located on chromosome 8 in humans (PMID: 8633055, 8698233). Until 2010, this locus was named plectin 1 (symbol PLEC1 in human; Plec1 in mouse and rat) and the gene product had been referred to as "hemidesmosomal protein 1" or "plectin 1, intermediate filament binding 500kDa". These names were superseded by plectin. The plectin gene locus in mouse on chromosome 15 has been analyzed in detail (PMID: 10556294, 14559777), revealing a genomic exon-intron organization with well over 40 exons spanning over 62 kb and an unusual 5' transcript complexity of plectin isoforms. Eleven exons (1-1j) have been identified that alternatively splice directly into a common exon 2 which is the first exon to encode plectin's highly conserved actin binding domain (ABD). Three additional exons (-1, 0a, and 0) splice into an alternative first coding exon (1c), and two additional exons (2alpha and 3alpha) are optionally spliced within the exons encoding the actin binding domain (exons 2-8). Analysis of the human locus has identified eight of the eleven alternative 5' exons found in mouse and rat (PMID: 14672974); exons 1i, 1j and 1h have not been confirmed in human. Furthermore, isoforms lacking the central rod domain encoded by exon 31 have been detected in mouse (PMID:10556294), rat (PMID: 9177781), and human (PMID: 11441066, 10780662, 20052759). The short alternative amino-terminal sequences encoded by the different first exons direct the targeting of the various isoforms to distinct subcellular locations (PMID: 14559777). As the expression of specific plectin isoforms was found to be dependent on cell type (tissue) and stage of development (PMID: 10556294, 12542521, 17389230) it appears that each cell type (tissue) contains a unique set (proportion and composition) of plectin isoforms, as if custom-made for specific requirements of the particular cells. Concordantly, individual isoforms were found to carry out distinct and specific functions (PMID: 14559777, 12542521,

18541706). In 1996, a number of groups reported that patients suffering from epidermolysis bullosa simplex with muscular dystrophy (EBS-MD) lacked plectin expression in skin and muscle tissues due to defects in the plectin gene (PMID: 8698233, 8941634, 8636409, 8894687, 8696340). Two other subtypes of plectin-related EBS have been described: EBS-pyloric atresia (PA) and EBS-Ogna. For reviews of plectin-related diseases see PMID: 15810881, 19945614. Mutations in the plectin gene related to human diseases should be named based on the position in NM_000445 (variant 1, isoform 1c), unless the mutation is located within one of the other alternative first exons, in which case the position in the respective Reference Sequence should be used. [provided by RefSeq, Aug 2011]

Immunogen	Recombinant protein of human Plectin
Gene ID	5339
Swiss Prot	Q15149
Synonyms	HD1; PCN; EBS1; EBSO; PLTN; EBSMD; EBSND; EBSOG; EBSPA; PLEC1; LGMD2Q; PLEC1b
Reactivity	Human, Mouse, Rat
Application	WB, IHC, IP
Recommended dilution	WB: 1:1000 IHC: 1:50 IP: 1:20
Calculated MW	532 kDa
Observed MW	400-500 kDa
Host species	Rabbit
Clonality	Polyclonal
Isotype	IgG
Purity	Affinity Purification
Conjugation	Un-conjugated
Storage Stability	Store at -20°C. Supplied in 50mM Tris-Glycine(pH 7.4), 0.15M NaCl, 40% Glycerol, 0.01% sodium azide and 0.05% BSA. Stable for 12 months from date of receipt.