

# PLEC1 Antibody (monoclonal) (M02)

531791

Mouse monoclonal antibody raised against a partial recombinant PLEC1. Catalog # AT3338a

### **Product Information**

**Application** WB, E **Primary Accession** Q15149 **Other Accession** NM 000445 Reactivity Human Host mouse Clonality monoclonal Isotype IgG2a Kappa **Clone Names** 4D12

#### **Additional Information**

Calculated MW

**Gene ID** 5339

Other Names Plectin, PCN, PLTN, Hemidesmosomal protein 1, HD1, Plectin-1, PLEC, PLEC1

Target/Specificity PLEC1 (NP\_000436, 4384 a.a. ~ 4493 a.a) partial recombinant protein with GST

tag. MW of the GST tag alone is 26 KDa.

**Dilution** WB~~1:500~1000 E~~N/A

**Format** Clear, colorless solution in phosphate buffered saline, pH 7.2.

**Storage** Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.

**Precautions** PLEC1 Antibody (monoclonal) (M02) is for research use only and not for use in

diagnostic or therapeutic procedures.

## Background

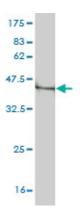
Plectin is a prominent member of an important family of structurally and in part functionally related proteins, termed plakins or cytolinkers, that are capable of interlinking different elements of the cytoskeleton. Plakins, with their multi-domain structure and enormous size, not only play crucial roles 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 (for reviews see 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 These names have been superseded by plectin.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 acting 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). It has been shown that 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.

### References

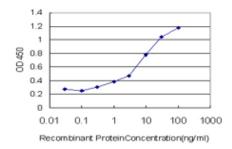
Proteome analysis of the thalamus and cerebrospinal fluid reveals glycolysis dysfunction and potential biomarkers candidates for schizophrenia. Martins-de-Souza D, et al. J Psychiatr Res, 2010 May 14. PMID 20471030. Epidermolysis bullosa simplex with muscular dystrophy. Chiav?rini C, et al. Dermatol Clin, 2010 Apr. PMID 20447487. PLEC1 mutations underlie adult-onset dilated cardiomyopathy in epidermolysis bullosa simplex with muscular dystrophy. Bolling MC, et al. J Invest Dermatol, 2010 Apr. PMID 20016501. Plectin gene defects lead to various forms of epidermolysis bullosa simplex. Rezniczek GA, et al. Dermatol Clin, 2010 Jan. PMID 19945614. BRCA2 interacts with the cytoskeletal linker protein plectin to form a complex controlling centrosome localization. Niwa T, et al. Cancer Sci, 2009 Nov. PMID 19709076.

### **Images**



Antibody Reactive Against Recombinant Protein.Western Blot detection against Immunogen (37.84 KDa).

Detection limit for recombinant GST tagged PLEC1 is approximately 0.3ng/ml as a capture antibody.



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