

Product datasheet for TA336512

MBNL1 Mouse Monoclonal Antibody [Clone ID: HL-1822 3A4-1E9]

Product data:

Product Type:	Primary Antibodies
Clone Name:	HL-1822 3A4-1E9
Applications:	WB
Recommend Dilution:	WB: 1:1000-1:5000, IF: 1:50-1:250, IHC: 1:1000, IHC-P: 1:1000
Reactivity:	Human, Mouse, Rat
Host:	Mouse
Isotype:	IgG1, kappa
Clonality:	Monoclonal
Immunogen:	Recombinant human Muscleblind-like 1 protein. [UniProt# Q9NR56]
Formulation:	Preservative: 0.1% Sodium Azide. Store at 4C short term. Aliquot and store at -20C long term. Avoid freeze-thaw cycles.
Concentration:	This product is unpurified. The exact concentration of antibody is not quantifiable.
Purification:	Tissue culture supernatant
Predicted Protein Size:	42 kDa
Gene Name:	muscleblind like splicing regulator 1
Database Link:	NP_066368 Entrez Gene 56758 MouseEntrez Gene 282635 RatEntrez Gene 4154 Human



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Background:

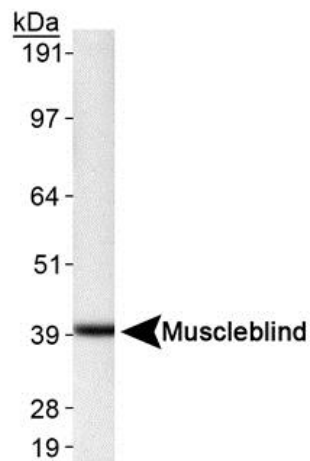
pre-mRNAs splicing /removal of non-coding regions is a critical process which contributes to a diverse proteome as well as gene expression regulation, and there are several splicing factors that are exclusively involved in a subset of splicing decisions - human muscleblind-like family of RNA-binding proteins: MBNL1/2/3 also known as MBNL/EXP, MBLL/MPL1, and MBXL/CHCR, respectively. Muscleblind-like 1 (MBNL1) is a known mediator of pre-mRNA alternative splicing regulation and it acts either as activator or repressor of splicing on specific pre-mRNA targets. Several pathologies are linked to expansion of polynucleotide sequences (mostly trinucleotide repeats) and CUG/CCUG expansion RNAs accumulates in Myotonic Dystrophy patient cells wherein it contributes to the disease by dysregulating alternative splicing factors such as CUGBP, Elav-like Family Member 1 (CELF1) and MBNL1. The expanded CUG repeats sequester MBNL proteins into nuclear foci, leading to loss of active protein and the sequestration of MBNL1 leads to missplicing of developmentally regulated events that are responsible for symptoms in DM types 1 and 2, such as the missplicing of the chloride channel (CLCN1) leading to myotonia. CELF1 and MBNL1 regulate a number of common splicing events, but in opposite directions and loss of MBNL1 activity is more critical than upregulation of CELF1 activity for abnormal splicing.

Synonyms:

EXP; EXP35; EXP40; EXP42; MBNL

Note:

This antibody is useful for Western blot where a band is seen at ~40 kDa, Immunofluorescence and Immunohistochemistry on paraffin-embedded sections.

Product images:

Western Blot: Muscleblind-like 1 Antibody (HL-1822 3A4-1E9) [TA336512] - Detection of Muscleblind in HeLa whole cell extract (33ug) using NB 110-37256.