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[Primary Antibody]

Rabbit Anti-MUSK antibody

Catalog Number: SL6473R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	MUSK
Immunogen:	KLH conjugated synthetic peptide derived from human MUSK: 231-330/869 <Extracellular>
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB=1:500-2000 IHC-P=1:100-500 IHC-F=1:100-500 ICC/IF=1:100-500 IF=1:50-200 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	(predicted: Human, Mouse, Rat, Dog, Pig, Cow)
Predicted MW.:	97kDa
Subcellular Location:	The cell membrane
Background:	Receptor tyrosine kinases (RTKs) represent an important class of transmembrane signaling molecules. Binding of the extracellular domain of an RTK to its cognate ligand leads to receptor dimerization and the activation of the intrinsic tyrosine kinase activity of its intracellular kinase domain. The specificity of this type of cellular communication is conferred in part by the distribution of the receptor, which determines the cells that are capable of responding to a given ligand. MuSK, for muscle-specific kinase, is an RTK that is uniquely specific to the skeletal muscle lineage. MuSK is expressed at low levels in proliferating myoblasts, but is induced upon terminal differentiation and myotube fusion. In

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	the embryo, MuSK is expressed in developing muscle, but its level of expression is dramatically reduced in mature muscle, where it is abundant only at the neuromuscular junction. The human MuSK gene maps to chromosome 9q31.3, overlapping a region containing the Fukuyama muscular dystrophy mutation. **Function:** Receptor tyrosine kinase which plays a central role in the formation and the maintenance of the neuromuscular junction (NMJ), the synapse between the motor neuron and the skeletal muscle. Recruitment of AGRIN by LRP4 to the MUSK signaling complex induces phosphorylation and activation of MUSK, the kinase of the complex. The activation of MUSK in myotubes regulates the formation of NMJs through the regulation of different processes including the specific expression of genes in subsynaptic nuclei, the reorganization of the actin cytoskeleton and the clustering of the acetylcholine receptors (AChR) in the postsynaptic membrane. May regulate AChR phosphorylation and clustering through activation of ABL1 and Src family kinases which in turn regulate MUSK. DVL1 and PAK1 that form a ternary complex with MUSK are also important for MUSK-dependent regulation of AChR clustering. May positively regulate Rho family GTPases through FNTA. Mediates the phosphorylation of FNTA which promotes prenylation, recruitment to membranes and activation of RAC1 a regulator of the actin cytoskeleton and of gene expression. Other effectors of the MUSK signaling include DNAJA3 which functions downstream of MUSK. May also play a role within the central nervous system by mediating cholinergic responses, synaptic plasticity and memory formation **Subunit:** Monomer (By similarity). Homodimer (Probable). Interacts with LRP4; the heterodimer forms an AGRIN receptor complex that binds AGRIN resulting in activation of MUSK (By similarity). Forms a heterotetramer composed of 2 DOK7 and 2 MUSK molecules which facilitates MUSK trans-autophosphorylation on tyrosine residue and activation. Interacts (via cytoplasmic part) with DOK7 (via IRS-type PTB domain); requires MUSK phosphorylation. Interacts with DVL1 (via DEP domain); the interaction is direct and mediates the formation of a DVL1, MUSK and PAK1 ternary complex involved in AChR clustering (By similarity). Interacts with PDZRN3; this interaction is enhanced by agrin (By similarity). Interacts with FNTA; the interaction is direct and mediates AGRIN-induced phosphorylation and activation of FNTA (By similarity). Interacts with CSNK2B; mediates regulation by CK2 (By similarity). Interacts (via the cytoplasmic domain) with DNAJA3 (By similarity). Interacts

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	with NSF; may regulate MUSK endocytosis and activity (By similarity). Interacts with CAV3; may regulate MUSK signaling (By similarity). Interacts with RNF31 (By similarity). **Subcellular Location:** Cell junction, synapse, postsynaptic cell membrane; Single-pass type I membrane protein (Probable). Note=Localizes to the postsynaptic cell membrane of the neuromuscular junction **Post-translational modifications:** Ubiquitinated by PDZRN3. Ubiquitination promotes endocytosis and lysosomal degradation (By similarity). Phosphorylated. Phosphorylation is induced by AGRIN. Autophosphorylation at Tyr-554 is required for interaction with DOK7 which in turn stimulates the phosphorylation and the activation of MUSK. DISEASE: Defects in MUSK is a cause of congenital myasthenic syndrome with acetylcholine receptor deficiency (CMS-ACHRD) [MIM:608931]. A postsynaptic congenital myasthenic syndrome. Mutations underlying AChR deficiency cause a 'loss of function' and show recessive inheritance. Note=MUSK mutations lead to decreased agrin-dependent AChR aggregation, a critical step in the formation of the neuromuscular junction. **Similarity:** Belongs to the protein kinase superfamily. Tyr protein kinase family. Contains 1 FZ (frizzled) domain. Contains 3 Ig-like C2-type (immunoglobulin-like) domains. Contains 1 protein kinase domain. **Database links:** **Entrez Gene:** 4593 Human **Entrez Gene:** 18198 Mouse **Entrez Gene:** 81725 Rat Omim: 601296 Human **SwissProt:** O15146 Human **SwissProt:** Q61006 Mouse **SwissProt:** Q62838 Rat **Unigene:** 521653 Human **Unigene:** 16148 Mouse **Unigene:** 10210 Rat

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