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MUSK Antibody

Purified Mouse Monoclonal Antibody
Catalog # AO1287a

Product Information

Application	IHC, ICC, E
Primary Accession	O15146
Reactivity	Human
Host	Mouse
Clonality	Monoclonal
Clone Names	10A4
Isotype	IgG1
Calculated MW	97056
Description	MuSK (for Muscle Specific Kinase) is a receptor tyrosine kinase required for the formation of the neuromuscular junction (NMJ). It induces cellular signaling by causing the addition of phosphate molecules to particular tyrosines on itself, and on proteins which bind the cytoplasmic domain of the receptor. It is activated by a nerve-derived proteoglycan called agrin. During development, the growing end of motor neuron axons secrete a protein called agrin.This protein binds to several receptors on the surface of skeletal muscle. The receptor which seems to be required for formation of the neuromuscular junction (NMJ), which comprises the nerve-muscle synapse is called MuSK. MUSK mutations lead to decreased agrin-dependent AchR aggregation, a critical step in the formation of the neuromuscular junction.
Immunogen	Purified recombinant extracellular fragment of human MUSK (aa24-209) fused with hIgGFc tag expressed in HEK293 cell line.
Formulation	Ascitic fluid containing 0.03% sodium azide.

Additional Information

Gene ID	4593
Other Names	Muscle, skeletal receptor tyrosine-protein kinase, 2.7.10.1, Muscle-specific tyrosine-protein kinase receptor, MuSK, Muscle-specific kinase receptor, MUSK
Dilution	IHC~~1/200 - 1/1000 ICC~~N/A E~~N/A
Storage	Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	MUSK Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	MUSK
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 (PubMed:25537362). 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 in acetylcholinesterase (AChE) localization at the neuromuscular junctions (NMJ) via its interaction wit COLQ (By similarity). May also play a role within the central nervous system by mediating cholinergic responses, synaptic plasticity and memory formation (By similarity).
Cellular Location	Postsynaptic cell membrane; Single-pass type I membrane protein. Note=Colocalizes with acetylcholine receptors (AChR) to the postsynaptic cell membrane of the neuromuscular junction

References

1. J Neuroimmunol. 2006 Aug;177(1-2):119-31. 2. Ann N Y Acad Sci. 2008;1132:76-83.

Images

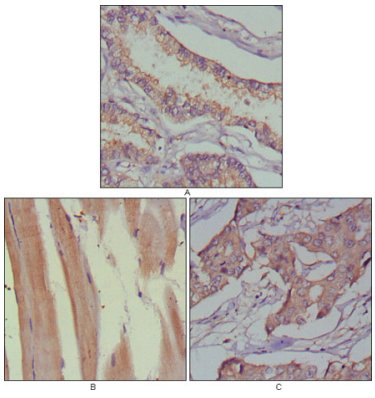
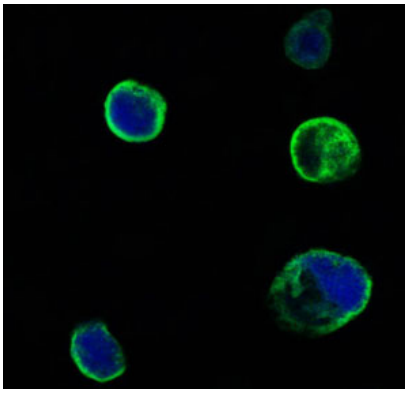


Figure 1: Immunohistochemical analysis of paraffin-embedded human lung cancer (A), muscles (B) and breast cancer (C) using MUSK mouse mAb with DAB staining.

Figure 2: Confocal immunofluorescence analysis of HEK293 cells trasfected with extracellular MUSK (aa24-209)-hIgGFc using MUSK mouse mAb (green). Blue: DRAQ5 fluorescent DNA dye.



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