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MUSK Antibody (N-term)

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP7664A

Product Information

Application	WB, FC, IHC-P, E
Primary Accession	O15146
Other Accession	Q62838, Q61006
Reactivity	Human
Predicted	Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	97056
Antigen Region	35-65

Additional Information

Gene ID	4593
Other Names	Muscle, skeletal receptor tyrosine-protein kinase, Muscle-specific tyrosine-protein kinase receptor, MuSK, Muscle-specific kinase receptor, MUSK
Target/Specificity	This MUSK antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 35-65 amino acids from the N-terminal region of human MUSK.
Dilution	WB~~1:1000 FC~~1:10~50 IHC-P~~1:100~500 E~~Use at an assay dependent concentration.
Format	Purified monoclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein G column, followed by dialysis against PBS.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	MUSK Antibody (N-term) 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

Background

Protein kinases are enzymes that transfer a phosphate group from a phosphate donor, generally the g phosphate of ATP, onto an acceptor amino acid in a substrate protein. By this basic mechanism, protein kinases mediate most of the signal transduction in eukaryotic cells, regulating cellular metabolism, transcription, cell cycle progression, cytoskeletal rearrangement and cell movement, apoptosis, and differentiation. With more than 500 gene products, the protein kinase family is one of the largest families of proteins in eukaryotes. The family has been classified in 8 major groups based on sequence comparison of their tyrosine (PTK) or serine/threonine (STK) kinase catalytic domains. The tyrosine kinase (TK) group is mainly involved in the regulation of cell-cell interactions such as differentiation, adhesion, motility and death. There are currently about 90 TK genes sequenced, 58 are of receptor protein TK (e.g. EGFR, EPH, FGFR, PDGFR, TRK, and VEGFR families), and 32 of cytosolic TK (e.g. ABL, FAK, JAK, and SRC families).

References

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Robinson D, et al. Oncogene 2000. 19: 5548.
Van der Ven, P, et al. Hum. Molec. Genet. 1993. 2: 1889.
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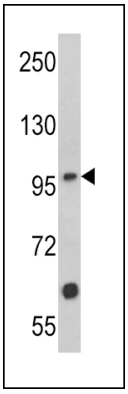
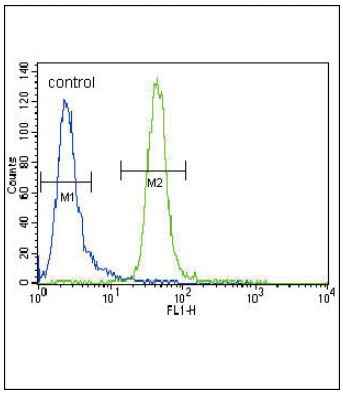
Van Weering D, et al. Recent Results Cancer Res. 1998. 154: 271.

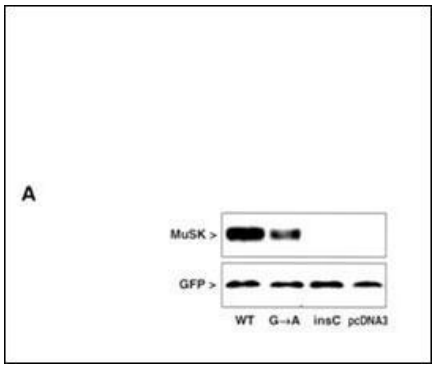
Images

Western blot analysis of MUSK Antibody (N-term) (Cat. #AP7664a) in Jurkat cell line lysates (35ug/lane). MUSK (arrow) was detected using the purified Pab.



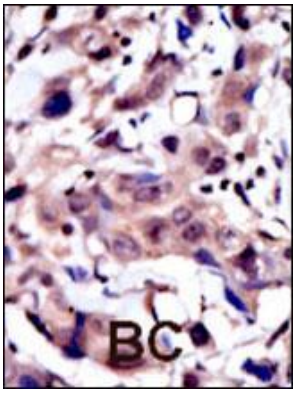


MUSK Antibody (N-term) (Cat. #AP7664a) flow cytometric analysis of CEM cells (right histogram) compared to a negative control cell (left histogram).FITC-conjugated goat-anti-rabbit secondary antibodies were used for the analysis.



MuSK protein expression in extracts of COS cells after transfection with MuSK mutated and GFP constructs. WB with polyclonal MuSK and monoclonal GFP antibodies showed normal expression of the wild-type MuSK protein (WT), diminished expression of the GA mutant MuSK and no expression of the insC mutant or the pcDNA3 vector alone in transfected COS cells. GFP cotransfection was used to verify transfection efficiency.

HA-tagged MuSK and GFP constructs were transfected into C2C12 cells. Transfected C2C12 cells were treated with cycloheximide (CHX) and at different times after addition of CHX, amounts of WT and mutant MuSK were analyzed by WB with anti-HA antibody. Alpha-tubulin was used as an internal control and transfection efficiency was verified with anti-GFP antibody.



Formalin-fixed and paraffin-embedded human cancer tissue reacted with the primary antibody, which was peroxidase-conjugated to the secondary antibody, followed by AEC staining. This data demonstrates the use of this antibody for immunohistochemistry; clinical relevance has not been evaluated. BC = breast carcinoma; HC = hepatocarcinoma.

Citations

- [Pathophysiological characterization of congenital myasthenic syndromes: the example of mutations in the MUSK gene].
- MUSK, a new target for mutations causing congenital myasthenic syndrome.