

Mouse anti-Myotilin, clone RSO34 (monoclonal)

Clone no. RSO34

MONXtra

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Product name	Mouse anti-Myotilin, clone RSO34 (monoclonal)
Host	Mouse
Applications	IHC-P (1:40-1:80), IHC-fr (1:20)
Species reactivity	human
Conjugate	-
Immunogen	Prokaryotic recombinant protein corresponding to a C-terminal region of 266 amino acids of the human myotilin molecule.
Isotype	IgG1, kappa
Clonality	Monoclonal
Clone number	RSO34
Size	1 ml
Concentration	n/a
Format	-
Storage buffer	Lyophilized
Storage until expiry date	2-8°C

FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES

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**Additional info**

Reconstitute with 1 mL or 0.1 mL of sterile distilled water as indicated on vial label. The myotilin gene on chromosome 5q31 encodes a 498 amino acid polypeptide with a molecular weight of 57kD. Myotilin is a structural protein of sarcomeric Z discs and sarcolemma in human skeletal and cardiac muscle. It is homologous to palladin and titin in the two C-terminal Ig-domains and also to palladin in its unique serine-rich N-terminal region. Myotilin interacts with alpha-actinin, actin and gamma-filamin. Mutations in the myotilin gene are associated with limb-girdle muscular dystrophy 1 A (LGMD1A) and one form of Myofibrillar Myopathy. It is highly conserved between human and mouse with its expression being more widespread in the embryo than in the adult. Expression of myotilin has been reported in adult skeletal and cardiac muscle with variable expression reported in the peripheral nervous system, lung, liver and kidney. NCL-MYOTILIN will be of use in studies to determine the expression of myotilin in normal and pathological tissues.

**References**

1. Mologni L et al. Mechanisms of Development. 103: 121–125 (2001)
2. Mykkänen OM et al. Mol. Biol. Cell. 12 (10): 3060–3073 (2001)
3. Hauser MA et al. Human Molecular Genetics. 9 (14): 2141–2147 (2000)
4. van der Ven PFM et al. Journal of Cell Biology. 151 (2): 235–247 (2000)
5. Salmikangas P et al. Human Molecular Genetics. 8 (7): 1329–1336 (1999)

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