

Product datasheet for **TA355325**

Myotilin (MYOT) Mouse Monoclonal Antibody [Clone ID: RSO34]

Product data:

Product Type:	Primary Antibodies
Clone Name:	RSO34
Applications:	IHC
Recommended Dilution:	1:20 (Frozen) 1:40 - 1:80 (Paraffin)
Reactivity:	Human
Host:	Mouse
Isotype:	IgG1
Clonality:	Monoclonal
Immunogen:	Prokaryotic recombinant protein corresponding to a C-terminal region of 266 amino acids of the human myotilin molecule
Specificity:	Human myotilin
Formulation:	Lyophilized tissue culture supernatant containing 15 mM sodium azide as a preservative.
Reconstitution Method:	The user is required to reconstitute the contents of the vial with the correct volume of sterile distilled water as indicated on the vial label
Conjugation:	Unconjugated
Storage:	Store at 2-8°C
Stability:	12 months
Gene Name:	myotilin
Database Link:	Entrez Gene 9499 Human Q9UBF9



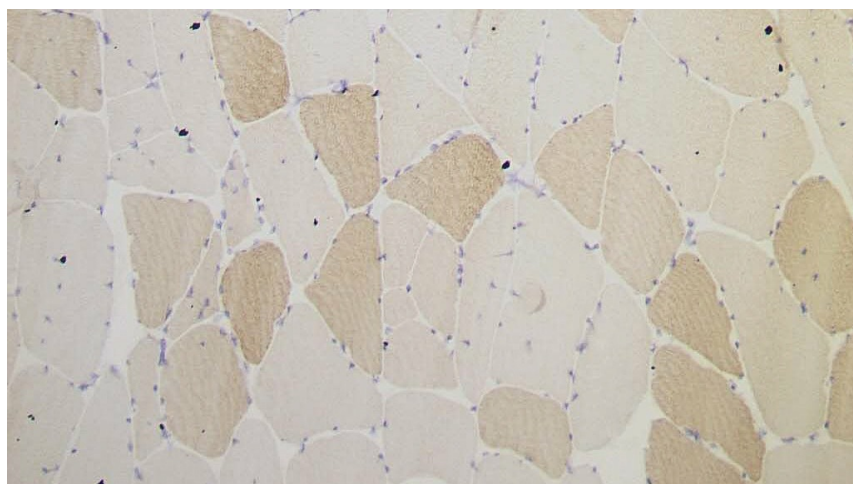
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Background:

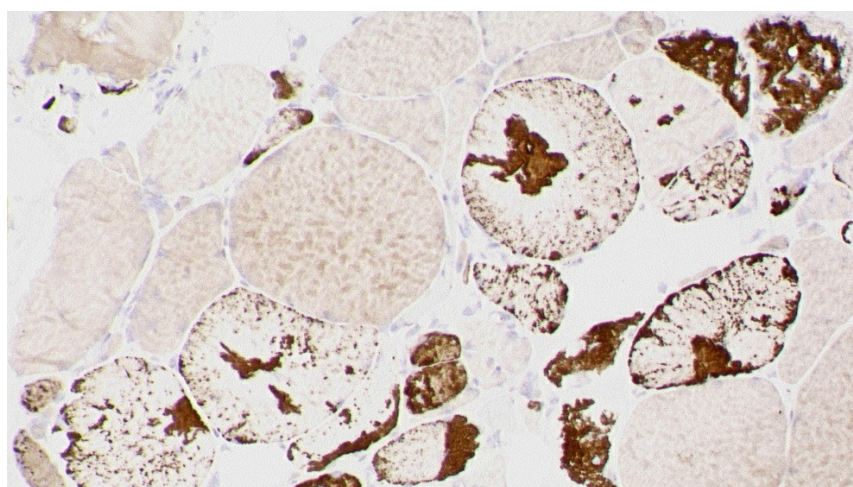
The myotilin gene on chromosome 5q31 encodes a 498 amino acid polypeptide with a molecular weight of 57kD. Myotilin is a structural protein of sarcomeric Z discs and sarcolemma in human skeletal and cardiac muscle. It is homologous to palladin and titin in the two C-terminal Ig-domains and also to palladin 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.

Synonyms:

LGMD1; LGMD1A; myotilin; TTID

Product images:

Human skeletal muscle: immunohistochemical staining for Myotilin. Note sarcoplasmic staining of normal muscle fibers (A) and presence of protein aggregates (B). Myotilin: clone RSO34



Human skeletal muscle: immunohistochemical staining for Myotilin. Note sarcoplasmic staining of normal muscle fibers (A) and presence of protein aggregates (B). Myotilin: clone RSO34