MUSCLE DISEASE ANTIBODIES

ADVANCING MUSCLE DISEASE DIAGNOSIS, MANAGEMENT AND RESEARCH

NOVOCASTRA
RESULTS YOU CAN RELY ON
Novocastra Muscle Disease Antibodies

The Novocastra muscle disease portfolio comprises 23 key clones for scientific research or in vitro diagnostic use, for use in IHC or western blotting. The majority of hybridomas are exclusive to Leica Biosystems, indicated by the **EXCLUSIVE** symbol in this catalog.

Proteins Involved in Muscular Dystrophy

Different types of muscular dystrophy can arise from mutations in numerous genes. Immunohistochemical stains may demonstrate alteration in expression or localization of the mutant protein, thus facilitating the diagnostic process. Direct or functional association between several of these proteins is reflected by the presence of specific secondary abnormalities. In particular, the expression of proteins of the dystrophin glycoprotein complex (DGC) appears to be strictly connected.

Dystrophin is a rod-like cytoskeletal protein which lies close to the muscle membrane and forms part of a system which links actin on the inside of muscle fibers, through a complex of trans-membrane proteins to extracellular matrix proteins which surround the muscle fibers. The dystrophin glycoprotein complex contains two groups of membrane proteins: alpha and beta-dystroglycans and alpha, beta, gamma and delta-sarcoglycans. The dystroglycans form a trans-membrane link between the cytoskeletal protein, dystrophin and the extracellular matrix proteins, such as laminin and agrin. The sarcoglycans form a group of trans-membrane proteins that are closely associated with the dystroglycans.

Mutations which lead to altered dystrophin expression cause Duchenne and Becker muscular dystrophy. Mutations in the gene for the muscle laminin alpha 2 (merosin) chain cause a form of congenital muscular dystrophy. In addition, four forms of limb girdle muscular dystrophy: LGMD2D, LGMD2E, LGMD2C and LGMD2F, are caused by mutations in the genes for alpha, beta, gamma and delta-sarcoglycans, respectively; therefore, many forms of muscular dystrophy are caused by defective expression of one or more of the component of the DGC and a differential diagnosis may be achieved by labeling muscle biopsies with antibodies to these proteins.

Proteins affected in muscular dystrophies may have diverse localization. Dysferlin is usually expressed at the sarcolemma and mutations in the DYSF gene cause LGMD2B and Miyoshi Myopathy. Mutations in genes encoding the intracellular proteins myotilin and calpain 3 cause LGMD1A (and myofibrillar myopathy) and LGMD2A, respectively. Emerin is a component of the nuclear envelope and mutations in the EMD gene are responsible for Emery-Dreifuss Muscular Dystrophy.

Labeling with an antibody to beta-spectrin, to monitor the quality of the sample (e.g. membrane integrity, protein degradation), is an essential immunohistochemical control.
# Product Details and Utility

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*Regulatory status in USA and EU. Regulatory status varies geographically. Please consult your Leica Biosystems representative for more information about regulatory classification in your country.
Beta-Dystroglycan **EXCLUSIVE**

Dystrophin associated glycoproteins (DAGs) are involved in the attachment of dystrophin to muscle membranes. The biological significance of this dystrophin/glycoprotein complex is not fully understood, but it appears to form an essential linkage between actin on the inside of the muscle fiber and muscle laminin in the basal lamina which surrounds the fiber. Beta-dystroglycan spans the muscle membrane and it has been suggested that it is the member of the complex which binds directly to dystrophin.

**1 mL Lyophilized Monoclonal (NCL-b-DG)**

For *in vitro* diagnostic use.

**Clone:** 43DAG1/8D5.

**Utility:** Qualitative identification by light microscopy of beta-dystroglycan by immunohistochemistry. Recommended for use on frozen sections.

**Immunogen:** Synthetic peptide containing 15 of the last 16 amino acids at the extreme C-terminus of the human beta-dystroglycan sequence (PKNMTYRSPPPYP-PCOOH).

**Specificity:** Human beta-dystroglycan (43 kD). Also cross-reacts strongly with beta-dystroglycan in sections of mouse, rat, rabbit, dog and chicken, hamster and toad muscle. Other animal species not tested.

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Calpain Antibodies **EXCLUSIVE**

The gene responsible for LGMD2A has been identified as the chromosome 15q15-encoded muscle-specific calcium-activated neutral protease, calpain 3. Calpain 3 enzyme is only stable in human muscle when homogenized in treatment buffer immediately after harvest (Anderson LVB et al. Am. J. of Pathol. 153(4), 1169-1179 (1998)), and in homogenates containing SDS and is therefore well suited for analysis by Western blot. NCL-CALP-2C4 reacts with the full-size calpain 3 (94kD) and an additional fragment (30kD) in human skeletal muscle. NCL-CALP-12A2 reacts with full-size protein plus apparent degradation products at approximately 60kD.

**2.5 mL Lyophilized Monoclonal (NCL-CALP-2C4)**

For research use only.

**Clone:** Calp3d/2C4.

**Utility:** Western blotting.

**Immunogen:** Synthetic peptide containing amino acids 1–19 of the human calpain 3 sequence.

**Specificity:** This antibody reacts with full-size calpain 3 (94 kD) plus an additional fragment at 30 kD in human skeletal muscle. A band of 94 kD is seen with rabbit and dog muscle while extracts of hamster muscle show reactivity with the 94 kD band and a larger species of approximately 110 kD. This 110 kD band is the principal immunoreactive species seen in rat muscle extracts. This antibody produces no bands with mouse, pig or chicken muscle.

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**2.5 mL Lyophilized Monoclonal (NCL-CALP-12A2)**

For research use only.

**Clone:** Calp3c/12A2.

**Utility:** Western blotting.

**Immunogen:** Synthetic peptide containing amino acids 355-370 of the human calpain 3 sequence.

**Specificity:** This antibody reacts with full-size calpain 3 (94 kD) plus an additional breakdown product at 60 kD in human skeletal muscle. The 94 kD band can be seen in muscle extracts from rabbit, mouse, dog, chicken, hamster, pig and rat. Degraded calpain 3 bands starting at approximately 60 kD are also usually present. Additional bands corresponding in size to calpains 1 and/or 2 can be detected in skeletal muscle from mouse, rat, chicken and hamster.
Dysferlin Antibodies EXCLUSIVE

Dysferlin is the protein product of the 2p13 gene that is defective in patients with Limb-Girdle Muscular Dystrophy type 2B (LGMD2B) and Miyoshi Myopathy (MM). Dysferlin is normally localized to the muscle plasma membrane. In patients with LGMD2B and MM, immunoreactivity to dysferlin is severely reduced or lost. Patients with other neuromuscular conditions demonstrate normal labeling patterns. NCL-Hamlet may require heat-induced epitope retrieval in some cases.

1 mL Lyophilized Monoclonal (NCL-Hamlet)
For *in vitro* diagnostic use.

**Clone:** Ham1 /7B6.

**Utility:** Qualitative identification by light microscopy of dysferlin by immunohistochemistry. Recommended for use on paraffin or frozen sections.

**Immunogen:** Synthetic peptide containing amino acids 1999-2016 of the human dysferlin molecule.

**Specificity:** Reactive with the dysferlin molecule in human skeletal muscle. Also present in many non-muscle tissues.

1 mL Lyophilized Monoclonal (NCL-Hamlet-2)
For *in vitro* diagnostic use.

**Clone:** Ham3/17B2.

**Utility:** Qualitative identification by light microscopy of dysferlin by immunohistochemistry. Recommended for use on paraffin or frozen sections.

**Immunogen:** Synthetic peptide containing amino acids 349-366, spanning exons 11 and 12, of the human dysferlin molecule.

**Specificity:** Reactive with the dysferlin molecule in human skeletal muscle. Also present in many non-muscle tissues.

Dystrophin Antibodies EXCLUSIVE

Duchenne Muscular dystrophy (DMD) is the most common of the muscular dystrophies resulting in progressive muscular wasting and death. Dystrophin is the 427kD protein product of the DMD gene located on the X chromosome at position Xp21. Abnormalities in protein expression occur in patients with DMD/BMD and dystrophin analysis may be used to distinguish these conditions from other neuromuscular diseases. Severe Duchenne muscular dystrophy is associated with a marked dystrophin deficiency, whereas patients with the milder form of Becker muscular dystrophy show less pronounced abnormalities of protein expression. The immunolabeling patterns for NCL-DYS1, NCL-DYS2 and NCL-DYS3 are similar; however, the use of all three antibodies is recommended to avoid the possibility of occasional false negative results.

Dystrophin (Rod Domain)

2.5 mL Lyophilized Monoclonal (NCL-DYS1)
For *in vitro* diagnostic use.

**Clone:** Dy4/6D3.

**Utility:** Qualitative identification by light microscopy of dystrophin (rod domain) by immunohistochemistry. Recommended for use on frozen sections.

**Immunogen:** Bacterial fusion protein (Hoffman EP et al., 1987).

**Specificity:** Reacts strongly with the rod domain (between amino acids 1181 and 1388) of human dystrophin. Also reacts with skeletal, cardiac and smooth muscle dystrophin from normal mouse, rat, rabbit, dog, hamster and pig. No reactivity with mdx mouse tissue or DMD/BMD patients who have a gene modification which removes the antibody binding site. No reaction is seen with chicken dystrophin.
**Dystrophin (C-terminus)**

**2.5 mL Lyophilized Monoclonal (NCL-DYS2)**
For *in vitro* diagnostic use.

- **Clone:** Dy8/6C5.
- **Utility:** Qualitative identification by light microscopy of dystrophin (C-terminus) by immunohistochemistry. Recommended for use on frozen sections.
- **Immunogen:** Synthetic polypeptide consisting of the last 17 amino acids at the carboxy terminus of the human dystrophin sequence.
- **Specificity:** Reacts strongly with the carboxy terminus (between amino acids 3669 and 3685) of human dystrophin. Also cross-reacts strongly with skeletal, cardiac and smooth muscle dystrophin from normal mouse, rat, rabbit, dog, chicken and hamster. No cross-reactivity with mdx mouse tissue. Cross-reacts very weakly with pig dystrophin.

**Dystrophin (N-terminus)**

**2.5 mL Lyophilized Monoclonal (NCL-DYS3)**
For *in vitro* diagnostic use.

- **Clone:** Dy10/12B2.
- **Utility:** Qualitative identification by light microscopy of dystrophin (N-terminus) by immunohistochemistry. Recommended for use on frozen sections.
- **Immunogen:** Fusion protein containing amino acids 67 to 713.
- **Specificity:** Reacts strongly with the amino terminal domain (between amino acids 321 and 494) of human dystrophin. Patient immunoreactivity indicates epitope is near exons 10 to 12. Epitope mapping suggests that sequences from amino acids 308 to 351 are involved in antibody binding. This region spans the junction of exons 9 and 10 and the epitope recognized may be part of a hinge region joining the amino domain to the central rod domain. No reactivity with DMD/BMD patients deleted for exons 10 to 12. No cross-reaction is observed with mouse (high background only), rat, rabbit, dog, chicken and hamster and pig dystrophin.

**Dystrophin**

NCL-DYSA is raised to an area of the dystrophin molecule, upstream from the C-terminal region and NCL-DYSB is raised to an area of the N-terminus of the dystrophin molecule. These two antibodies will be of particular interest in the investigation of archived formalin-fixed, paraffin-embedded material.

- **1 mL Lyophilized Monoclonal (NCL-DYSA)**
  - **Clone:** 13H6.
  - **Utility:** Immunohistochemistry. Recommended for use on paraffin sections.
  - **Immunogen:** Prokaryotic recombinant protein corresponding to a region of the rod domain of the human dystrophin molecule.
  - **Specificity:** Human dystrophin molecule.

- **1 mL Lyophilized Monoclonal (NCL-DYSB)**
  - **Clone:** 34C5.
  - **Utility:** Immunohistochemistry. Recommended for use on paraffin sections.
  - **Immunogen:** Prokaryotic recombinant protein corresponding to amino acids 321 to 494 of the dystrophin molecule.
  - **Specificity:** Human dystrophin molecule.
Emerin EXCLUSIVE

Emery-Dreifuss muscular dystrophy (EDMD) is a late onset, X-linked, recessive disorder characterized by slowly progressing contractures, wasting of skeletal muscle and cardiomyopathy usually presented as heart block. Contractures are seen in the elbows, Achilles tendons and postcervical muscles with humero-peroneal distribution early in the course of the disease. The STA gene, at Xq28 locus, encodes a serine-rich 34kD protein, emerin, which is ubiquitous in tissues and is found in highest concentration in skeletal and cardiac muscle. Emerin is localized in the nuclear membrane of normal muscle cells and its deficiency plays a crucial part in the pathology of EDMD.

1 mL Lyophilized Monoclonal (NCL-EMERIN)
For in vitro diagnostic use.

Clone: 4G5.
Utility: For the qualitative identification by light microscopy of emerin by immunohistochemistry. Recommended for use on paraffin or frozen sections.
Immunogen: Prokaryotic recombinant protein corresponding to a 222 amino acid region near the N-terminus of the emerin protein.
Specificity: Human emerin protein.

Merosin Laminin Alpha 2 Chain EXCLUSIVE

The muscle-specific form of laminin, merosin, is composed of three chains: alpha 2, beta 1 and gamma 1.

Mutations in the chromosome 6 encoded gene for the laminin alpha 2 chain of merosin are responsible for a form of congenital muscular dystrophy (CMD). Merosin-negative CMD is characterized by a severe clinical phenotype and is associated with white matter changes on brain imaging.

1 mL Lyophilized Monoclonal (NCL-MEROSIN)
For in vitro diagnostic use.

Utility: Qualitative identification by light microscopy of merosin laminin alpha 2 chain by immunohistochemistry. Recommended for use on frozen sections.
Immunogen: Purified protein from placenta.
Specificity: Reacts strongly with laminin alpha 2 chain of merosin in human and rabbit skeletal muscle. No reaction is observed in muscle sections from mouse, rat, dog, chicken, hamster or pig.
Myosin Heavy Chain Antibodies

Myosin is a contractile muscle specific protein composed of two heavy and four light chains. The myosin heavy chain has many isoforms which are specific for different muscles or fiber types, some of which are developmentally regulated. The range of myosin heavy chain antibodies may prove useful for investigating development of intrafusal and extrafusal muscle fibers and the course of muscle fiber regeneration. At the ultrastructural level, antibodies can reveal architectural details of the myofilament as well as the cytoplasmic and membrane sites of new myosin integration.

Myosin Heavy Chain (developmental) EXCLUSIVE

1 mL Lyophilized Monoclonal (NCL-MHCd)
For research use only.

Clone: RNMy2/9D2.
Utility: Immunohistochemistry. Recommended for use on frozen sections.
Immunogen: Native myosin extracted from the hind limb muscle of 7 day old rats.
Specificity: Human myosin developmental type heavy chain. Note that this antibody recognizes a myosin heavy chain (MHC) present during the embryonic and neonatal period in the development of skeletal muscle. The same MHC occurs during regeneration of muscle fibers. Also reacts with rat myosin developmental type heavy chain.

Myosin Heavy Chain (fast)

1 mL Lyophilized Monoclonal (NCL-MHCf)
For research use only.

Clone: WB-MHCf.
Utility: Immunohistochemistry. Recommended for use on frozen sections.
Immunogen: Native myosin extracted from rabbit psoas muscle.
Specificity: Rabbit myosin fast type heavy chain. Cross-reacts with human myosin fast type heavy chain. The antibody also reacts with type II myosin heavy chain (both IIa and IIb) in rat, mouse, dog, sheep, pig and goat muscle.

Myosin Heavy Chain (neonatal)

1 mL Lyophilized Monoclonal (NCL-MHCn)
For research use only.

Clone: WB-MHCn.
Utility: Immunohistochemistry. Recommended for use on frozen sections.
Immunogen: Myosin extracted from the hind limb muscle of a 3 day old rabbit, denatured with sodium dodecyl sulphate.
Specificity: Rabbit myosin neonatal type heavy chain. Cross-reacts with human myosin neonatal type heavy chain. Note that this antibody recognizes a myosin heavy chain present during the neonatal period in rabbit limb muscle. The temporal appearance of an equivalent epitope may differ in different species and consequently it may not be correct to label the epitope as “neonatal” in some circumstances.

Myosin Heavy Chain (slow)

1 mL Lyophilized Monoclonal (NCL-MHCs)
For research use only.

Clone: WB-MHCs.
Utility: Immunohistochemistry. Recommended for use on frozen sections.
Immunogen: Native myosin extracted from rabbit soleus muscle.
Specificity: Rabbit myosin slow type heavy chain. Cross-reacts with human myosin slow type heavy chain. The antibody also reacts with type I myosin heavy chain in rat, mouse, dog, sheep, pig and goat muscle.
Myotilin **EXCLUSIVE**

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.

1 mL Lyophilized Monoclonal (NCL-MYOTILIN)
For research use only.

**Clone:** RSO34.

**Utility:** Immunohistochemistry. Recommended for use on paraffin and frozen sections.

**Immunogen:** Prokaryotic recombinant protein corresponding to a C-terminal region of 266 amino acids of the human myotilin molecule.

**Specificity:** Human myotilin.

Sarcoglycan Antibodies **EXCLUSIVE**

In normal skeletal muscle, dystrophin, the protein product of the gene which is defective in Duchenne and Becker muscular dystrophy, is attached to the muscle membrane via a complex of proteins (dystrophin-associated glycoproteins, DAG’s). Dystrophin-deficient muscle shows a generalized reduction in DAG labeling. The expression of different members of the dystrophin glycoprotein complex is altered in several types of muscular dystrophy. For example, patients with LGMD2D have mutations in the gene for alpha-sarcoglycan, those with LGMD2E have mutations in the beta-sarcoglycan gene, those with LGMD2C have mutations in the gamma-sarcoglycan gene and those with LGMD2F have mutations in the delta-sarcoglycan gene. As the sarcoglycans function together as a sub-complex, mutations in any one of the sarcoglycan genes usually results in variable expression for the whole group.

1 mL Alpha-Sarcoglycan (Adhalin) Liquid Monoclonal (NCL-L-a-SARC)
For *in vitro* diagnostic use.

**Clone:** Ad1/20A6.

**Utility:** NCL-L-a-SARC is intended for the qualitative identification by light microscopy of alpha-sarcoglycan (adhalin) by immunohistochemistry. Recommended for use on frozen sections.

**Immunogen:** Fusion protein containing amino acids 217 to 289 of the rabbit adhalin sequence (Roberds SL et al., 1993).

**Specificity:** Human alpha-sarcoglycan, also known as adhalin. Also cross-reacts strongly with alpha-sarcoglycan in sections of muscle from mouse, rat, rabbit, hamster and pig. Does not react with chicken muscle.

1 mL Beta-Sarcoglycan Liquid Monoclonal (NCL-L-b-SARC)
For *in vitro* diagnostic use.

**Clone:** βSarc1/5B1.

**Utility:** NCL-L-b-SARC is intended for the qualitative identification by light microscopy of beta-sarcoglycan by immunohistochemistry. Recommended for use on frozen sections.

**Immunogen:** Fusion protein RBSG-NT of the human beta-sarcoglycan sequence.

**Specificity:** Human beta-sarcoglycan (43 kD).
### Sarcoglycan Antibodies EXCLUSIVE

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<td>Immunogen:</td>
<td>Synthetic peptide containing amino acids 1-19 at the N-terminus of the human delta-sarcoglycan sequence (Jung D. et al., 1966).</td>
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<td>Specificity:</td>
<td>Human delta-sarcoglycan (35 kD). Does not react with delta-sarcoglycan in sections of mouse, rat, rabbit, dog, chicken, hamster or pig muscle. Other animal species not tested.</td>
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<td>Immunogen:</td>
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</tr>
<tr>
<td>Specificity:</td>
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Spectrin **EXCLUSIVE**

Spectrin is a cytoskeletal protein which has some structural homology with dystrophin, the protein that is defective in Duchenne and Becker muscular dystrophy. Subtle membrane damage frequently occurs during the excision and freezing of muscle biopsies. Labeling for spectrin must be used to monitor membrane integrity. NCL-SPEC1 recognizes the beta chain of spectrin in erythrocytes and muscle. NCL-SPEC1 reacts with human beta-spectrin.

**1 mL Lyophilized Monoclonal (NCL-SPEC1)**

For *in vitro* diagnostic use.

**Clone:** RBC2/3D5.

**Utility:** NCL-SPEC1 is intended for the qualitative identification by light microscopy of spectrin by immunohistochemistry. Recommended for use on frozen sections.

**Immunogen:** Human red blood cell membrane “ghosts”.

**Specificity:** Beta chain of spectrin in human red blood cells and muscle.

Urophin **EXCLUSIVE**

The utrophin gene is located on chromosome 6. The protein is a homologue of dystrophin and is known as dystrophin-related protein. In normal muscle, utrophin is restricted to neuromuscular junctions; however, in dystrophin-deficient muscle, utrophin expression may be upregulated and labeling appears around the periphery of most fibers. Immunohistochemical staining with NCL-DRP2 labels vessels and neuromuscular junctions and the upregulated form of utrophin, located around fiber membranes.

**2.5 mL Lyophilized Monoclonal (NCL-DRP2)**

For *in vitro* diagnostic use.

**Clone:** DRP3/20C5.

**Utility:** NCL-DRP2 is intended for the qualitative identification by light microscopy of utrophin (N-terminus) by immunohistochemistry. Recommended for use on frozen sections.

**Immunogen:** Fusion protein containing the first 261 amino acids of the published UTRN gene sequence.

**Specificity:** Amino terminal domain of the human homolog of human dystrophin, utrophin (also known as dystrophin-related protein or “DRP”). Also cross-reacts with utrophin in sections of muscle from rat and dog. Other animal species have not been tested.

Human skeletal muscle: immunohistochemical staining for utrophin using NCL-DRP2 antibody. In control muscle the antibody labels blood vessels and neuromuscular junctions (A). Urophin is expressed at the sarcolemma in individuals with mutations in the DMD gene (B). Frozen sections. Photographs supplied courtesy of Dr Rita Barresi.
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