MYOMEDIX

Datasheet: NEB-Nterm

Description: Rabbit polyclonal Antibody (batch#6969)
Specificity: Nebulin N-terminal 25kDa region
Other names: NEB-N rab ab
Product Type: Polyclonal Antibody
Isotype: Polyclonal IgG
Quantity: 100µL (40 µg)

Product Details

This antiserum has been raised to the amino-terminal 25kDa region of nebulin. This part of nebulin is not composed of highly repetitive sequences (1,2). It therefore provides a suitable region for specific epitope recognition. Structurally, this epitope localizes at the pointed-end region of the thin filament (3; see also Figure 2). Functionally, this segment of nebulin has been implicated in tropomodulin binding (4).
The NEB-Nterm antibody has been affinity-purified with the specific antigen coupled to an affinity resign. Affinity-purified IgGs after elution are provided at a concentration of 0.4 µg/µl. For long-term storage, aliquoting, snap-freezing and storage at -80°C is recommended. For storage for up to 6 months, storage at 4°C without re-freezing is recommended.
The specificity of this antibody has been verified by Western blots: Heart muscle does not express nebulin and instead a nebulin-homologous 107kDa protein called “nebulette” (5, 6). Specificity of Neb-Nterm in Western blots is indicated by reactivity to the 800kDa nebulin protein in skeletal muscle and lack of reactivity to heart muscle extracts (Figure 1, left).
Specificity has also been confirmed by comparing wildtype and Nebulin-KO mice: Inactivation of the nebulin gene in mice by gene targeting leads to nebulin-deficient myofibrils (7 8). Western blots comparing NEB-KO and WT mice confirms lack of nebulin expression in NEB-KO mice (Figure 1, right).
In human, nebulin-deficiency is causative for the main-stream form nebulin myopathy (9). Therefore, nebulin antibodies are also useful to detect nebulin deficiency (10) or truncations (11, 12) in patients with nemaline myopathies.

Rabbit IgG polyclonal NEB-Nterm:

Recommended usages: Western blots, immunofluorescence
Tested species: Human and mouse
Related Products: Nebulin-Cterm antibody, Nebulin-Cterm antibody (phospho-specific)

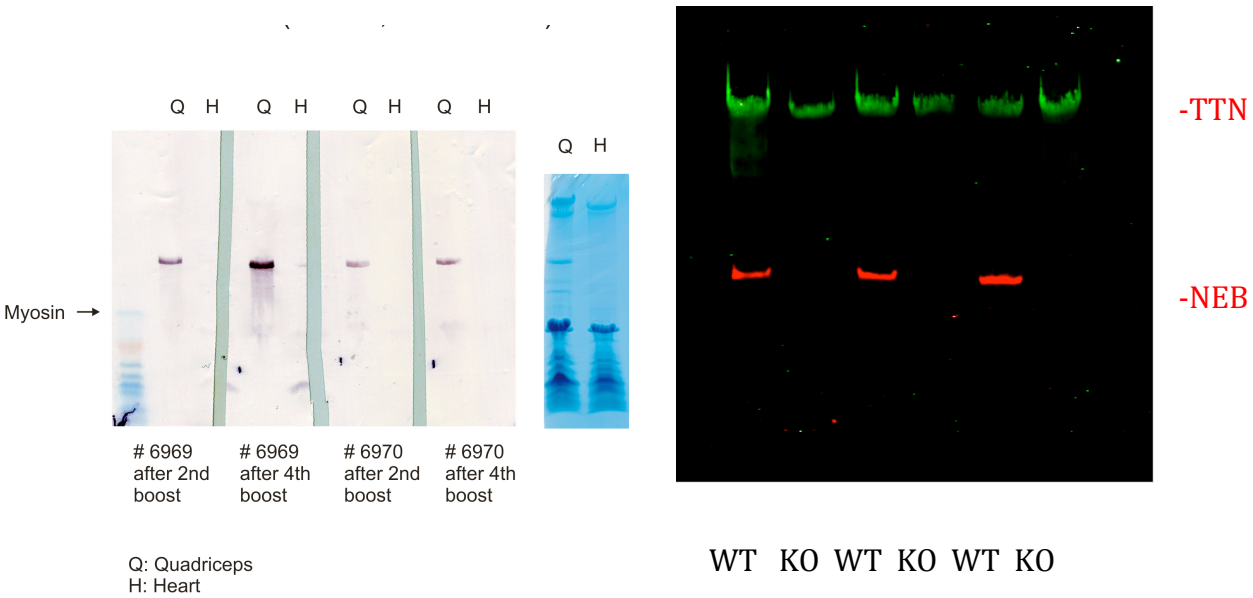

Figure 1. Western blot characaterizaion of anti-NEB-Nterm antibodies.
Left: In blots of skeletal muscle extracts from M. quadriceps, NEB-Nterm detects the 800kDa nebulin species, whereas heart extracts do not react.
Right: Simultaneous detection of titin and nebulin in the two channel-mode using different titin and nebulin antibodies (see also 8). Anti-NEB-Nterm detects nebulin in wild-type but not nebulin KO mice (Figure kindly provided by Danielle Buck and Henk Granzier, University of Arizona)

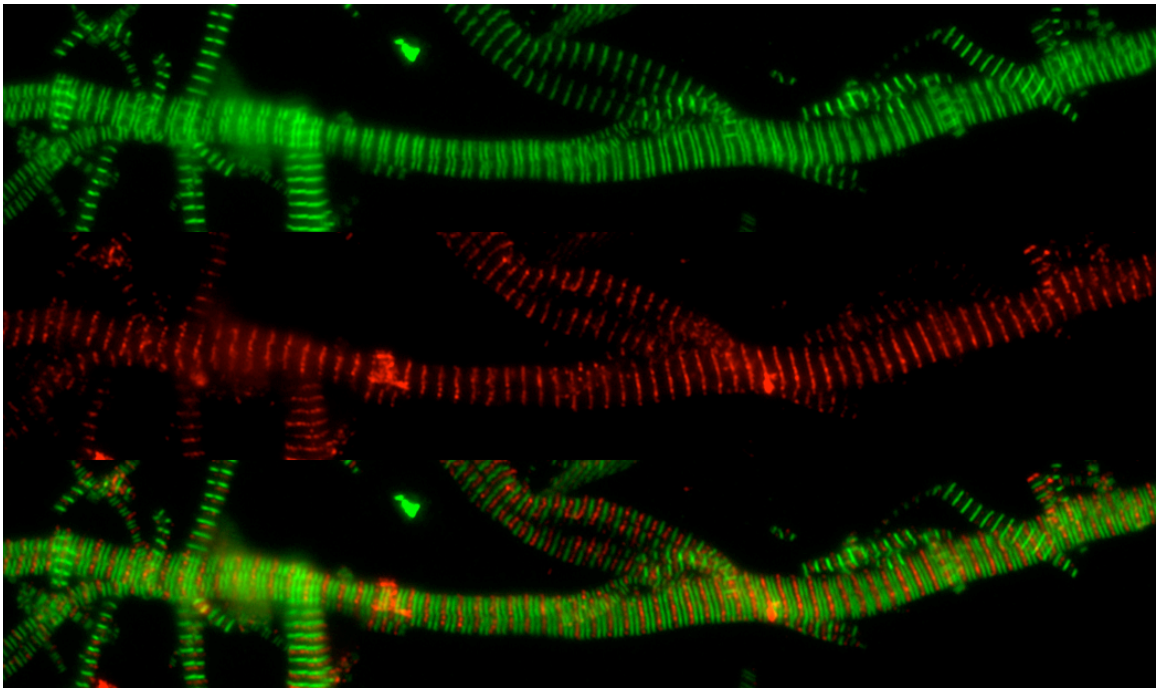

Figure 2. Immunofluorescence labeling of isolated myofibrils with Neb-Nterm (top; green) detects a doublet in each sarcomere. Double-labeling of Z-disks with desmin (below, red) assigns this doublet to project about 1 µm away from the Z-disk, thus consistent with labeling of thin filament pointed-ends (bottom: merge of green and red channels; courtesy of Akira Hanashima, Chiba University, Japan).

Literature

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MYOMEDIX GMBH

Myomedix GmbH ▪ MIttelstr. 42 ▪ 68169 Mannheim

Tel.: +49621 3832716 ▪ Email: info@myomedix.com

Web: www.myomedix.com

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