



Datasheet: NEB-Cterm

Description: Rabbit polyclonal Antibody (batch#6964)
Specificity: Nebulin C-terminal 35kDa region
Other names: NEB-C rab ab
Product Type: Polyclonal Antibody
Isotype: Polyclonal IgG
Quantity: 100µL (70 µg)

Product Details

This antiserum has been raised to the carboxy-terminal 35kDa 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 inside Z-disks within the barbed-end region of the thin filament (3). Functionally, this segment of nebulin has been implicated in binding to titin (4), to myopalladin (5), and to N-WASP after stimulation with IGF-1 (see (6) and related product: Nebulin Cterm – phospho-specific antibody).

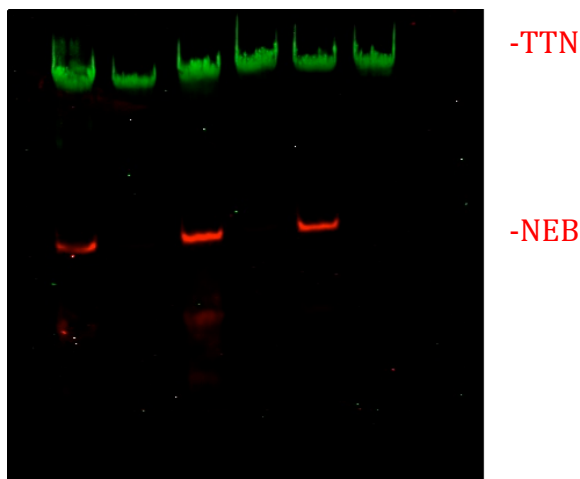
The NEB-Cterm antibody has been affinity-purified with the specific antigen coupled to an affinity resin. Affinity-purified IgGs after elution are provided at a concentration of 0.7 µg/µl. For long-term storage, aliquoting, snap-freezing and storage at -80°C is recommended. For storage for up to 6 months, storage at 4°C without re-freezing is recommended.

The specificity of this antibody has been verified by Western blots 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 with NEB-Cterm antibody confirm lack of nebulin expression in NEB-KO mice (Figure 1, right).

In human, nebulin-deficiency is causative for the mainstream form nebulin myopathy (9). Therefore, nebulin Cterm antibodies are useful to detect nebulin deficiency (or truncations (10-12) in patients with nemaline myopathies.

Rabbit IgG polyclonal NEB-Cterm:

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



WT KO WT KO WT KO

Figure 1. Western blot characterization of anti-NEB-Cterm antibodies. Simultaneous detection of titin and nebulin in the two channel-mode using different titin and nebulin antibodies (see also 8). Anti-NEB-Cterm detects nebulin in wild-type but not nebulin KO mice (Figure kindly provided by Danielle Buck and Henk Granzier, University of Arizona)

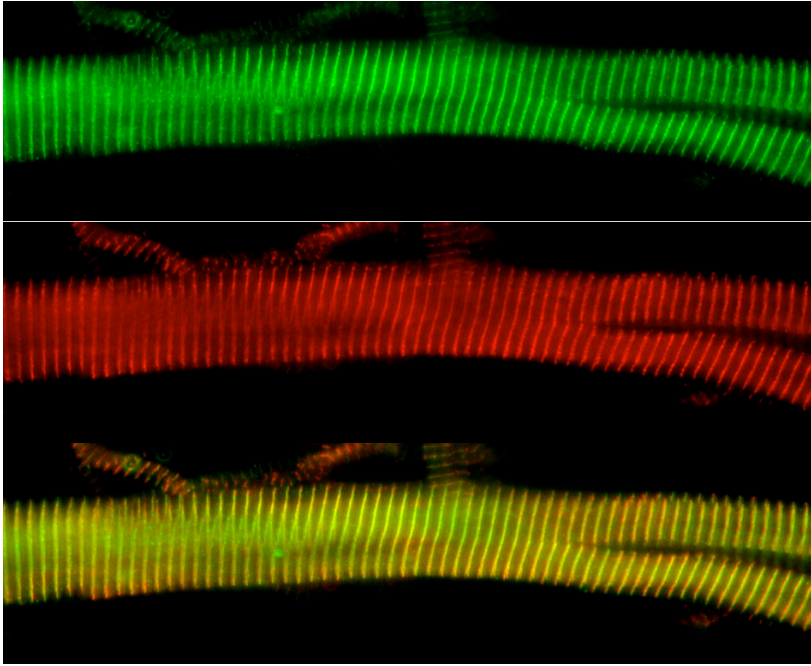


Figure 2.

Immunofluorescence labeling of isolated myofibrils with NEB-Cterm antibody (top; green).

Middle: Labeling of sarcomeric Z-disks with desmin (center, in red). The merge of both channels assigns the staining of NEB-Cterm to the sarcomeric Z-disk (bottom: merge in yellow; courtesy of Akira Hanashima, Chiba University, Japan).

Literature

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