Mouse Anti human dystrophin Hybridoma [NBOIJOHF3C Clone ]

Anti human dystrophin Hybridoma
Lot. No. (See product label)

CELL LINE INFORMATION

<table>
<thead>
<tr>
<th>Cat.No.</th>
<th>CSC-H2670</th>
</tr>
</thead>
<tbody>
<tr>
<td>Common Name</td>
<td>DMD</td>
</tr>
<tr>
<td>Clone</td>
<td>NBOIJOHF3C Clone</td>
</tr>
</tbody>
</table>

Cell Line Description

The antibody is against human dystrophin (Name: peptide AA684-699TVTTVTTREQILVKHA conjugated to BSA; Origin: synthetic human sequence; Chemical Composition: peptide; Developmental Stage: adult; Amino acid sequence analysis: AA684-699TVTTVTTREQILVKHA of human dystrophin)

Background

The dystrophin gene is the largest gene found in nature, measuring 2.4 Mb. The gene was identified through a positional cloning approach, targeted at the isolation of the gene responsible for Duchenne (DMD) and Becker (BMD) Muscular Dystrophies. DMD is a recessive, fatal, X-linked disorder occurring at a frequency of about 1 in 3,500 new-born males. BMD is a milder allelic form. In general, DMD patients carry mutations which cause premature translation termination (nonsense or frame shift mutations), while in BMD patients dystrophin is reduced either in molecular weight (derived from in-frame deletions) or in expression level. The dystrophin gene is highly complex, containing at least eight independent, tissue-specific promoters and two polyA-addition sites. Furthermore, dystrophin RNA is differentially spliced, producing a range of different transcripts, encoding a large set of protein isoforms. Dystrophin (as encoded by the Dp427 transcripts) is a large, rod-like cytoskeletal protein which is found at the inner surface of muscle fibers. Dystrophin is part of the dystrophin-glycoprotein complex (DGC), which bridges the inner cytoskeleton (F-actin) and the extra-cellular matrix.

Immunogen

human dystrophin

Immunological Donor
Balb/c Mouse spleen

Myeloma
Mouse Sp2/0

Fusion Species
Mouse X Mouse Hybridoma

Mycoplasma
Mycoplasma Status: Negative (MycoAlert Kit)

ANTIBODY INFORMATION

Reactivity
human (not dog)

Isotype
IgG1

Target
DMD

Application
Cell binding: muscle; Immunohistology: sarcolemma; Immunoblotting; Immunohistochemistry

SAFETY AND PACKAGING

Storage
liquid nitrogen

Safety Considerations
The following safety precautions should be observed.
1. Use pipette aids to prevent ingestion and keep aerosols down to a minimum.
2. No eating, drinking or smoking while handling the hybridoma.
3. Wash hands after handling the hybridoma and before leaving the lab.
4. Decontaminate work surface with disinfectant or 70% ethanol before and after working with hybridoma.
5. All waste should be considered hazardous.
6. Dispose of all liquid waste after each experiment and treat with bleach.
**ANTIGEN GENE INFORMATION**

<table>
<thead>
<tr>
<th><strong>Gene Name</strong></th>
<th><strong>DMD dystrophin [ Homo sapiens ]</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Official Symbol</strong></td>
<td>DMD</td>
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<tr>
<td><strong>Synonyms</strong></td>
<td>DMD;dystrophin;dystrophin (muscular dystrophy, Duchenne and Becker types), includes DXS142, DXS164, DXS206, DXS230, DXS239, DXS268, DXS269, DXS270, DXS272,BMD;DXS142;DXS164;DXS206;DXS230;DXS239;DXS268;DXS269;DXS270;DXS272;muscular dystrophy; Duchene and Becker types;CMD3B;NP_000100;NM_000109;P11532;OTTHUMP00000023117;OTTHUMP00000023121;OTTHUMP00000023124;OTTHUMP00000023125;OTTHUMP00000023126;OTTHUMP00000215590;OTTHUMP00000215591;OTTHUMP00000215592;OTTHUMP00000215846;HGNC: 2928;Entrez Gene: 1756;Ensembl: ENSG00000198947;OMIM: 300377;UniProtKB: P11532;DMD_HUMAN;</td>
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<tr>
<td><strong>Gene ID</strong></td>
<td>1756</td>
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<tr>
<td><strong>mRNA Refseq</strong></td>
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<td><strong>Protein Refseq</strong></td>
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<td><strong>MIM</strong></td>
<td>300377</td>
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<td><strong>UniProt ID</strong></td>
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<tr>
<td><strong>Chromosome Location</strong></td>
<td>Xp21.2</td>
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<tr>
<td><strong>Pathway</strong></td>
<td>Arrhythmogenic right ventricular cardiomyopathy (ARVC), organism-specific biosystem; Arrhythmogenic right ventricular cardiomyopathy (ARVC), conserved biosystem; Dilated cardiomyopathy, organism-specific biosystem; Dilated cardiomyopathy, conserved biosystem; Hypertrophic cardiomyopathy (HCM), organism-specific biosystem; Hypertrophic cardiomyopathy (HCM), conserved biosystem; Muscle contraction, organism-specific biosystem;</td>
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<tr>
<td><strong>Function</strong></td>
<td>PDZ domain binding; actin binding; actin binding; beta-dystroglycan binding; dystroglycan binding; integrin binding; metal ion binding; nitric-oxide synthase binding; protein binding; structural constituent of cytoskeleton; structural constituent of muscle; structural constituent of muscle; zinc ion binding;</td>
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</tbody>
</table>

**REFERENCES**