Product datasheet

Anti-Dystrophin antibody [MANDRA1] ab7164

Overview

Product name
Anti-Dystrophin antibody [MANDRA1]

Description
Mouse monoclonal [MANDRA1] to Dystrophin

Host species
Mouse

Tested applications
Suitable for: ELISA, IHC-Fr, WB, ICC/IF
Unsuitable for: IHC-P

Species reactivity
Reacts with: Mouse, Rat, Human, Fish

Immunogen
Fusion protein, corresponding to amino acids 3200-3684 of Human Dystrophin.

Epitope
128 amino acids at the end of the C-terminal domain of the human dystrophin molecule (a.a. residues 3558-3684).

Positive control
lympho blastoid cells, cultures of brain astroglial and neuronal cells, liver and Hep G2 cells

General notes
The C-terminal domain of the human dystrophin molecule (a.a. residues 3558-3684) is present in normal muscle tissue. It is also present in nearly all Becker muscular dystrophies, but is absent in cases of Duchenne muscular dystrophies and in the dystrophic mouse (mdx).

This product was changed from ascites to tissue culture supernatant on 17 May 2019. Please note that the dilutions may need to be adjusted accordingly. If you have any questions, please do not hesitate to contact our scientific support team.

Properties

Form
Liquid

Storage instructions
Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

Storage buffer
Preservative: 0.097% Sodium azide

Purity
Tissue culture supernatant

Primary antibody notes
The C-terminal domain of the human dystrophin molecule (a.a. residues 3558-3684) is present in normal muscle tissue. It is also present in nearly all Becker muscular dystrophies, but is absent in cases of Duchenne muscular dystrophies and in the dystrophic mouse (mdx).

Clonality
Monoclonal

Clone number
MANDRA1
Isotype
IgG1

Applications

Our Abpromise guarantee covers the use of ab7164 in the following tested applications. The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

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Application notes
Is unsuitable for IHC-P.

Target

Function
Anchors the extracellular matrix to the cytoskeleton via F-actin. Ligand for dystroglycan.
Component of the dystrophin-associated glycoprotein complex which accumulates at the neuromuscular junction (NMJ) and at a variety of synapses in the peripheral and central nervous systems and has a structural function in stabilizing the sarcolemma. Also implicated in signaling events and synaptic transmission.

Tissue specificity
Expressed in muscle fibers accumulating in the costameres of myoplasm at the sarcolemma.
Expressed in brain, muscle, kidney, lung and testis. Isoform 5 is expressed in heart, brain, liver, testis and hepatoma cells. Most tissues contain transcripts of multiple isoforms, however only isoform 5 is detected in heart and liver.

Involvement in disease
Defects in DMD are the cause of Duchenne muscular dystrophy (DMD) [MIM:310200]. DMD is the most common form of muscular dystrophy; a sex-linked recessive disorder. It typically presents in boys aged 3 to 7 year as proximal muscle weakness causing waddling gait, toe-walking, lordosis, frequent falls, and difficulty in standing up and climbing up stairs. The pelvic girdle is affected first, then the shoulder girdle. Progression is steady and most patients are confined to a wheelchair by age of 10 or 12. Flexion contractures and scoliosis ultimately occur. About 50% of patients have a lower IQ than their genetic expectations would suggest. There is no treatment.
Defects in DMD are the cause of Becker muscular dystrophy (BMD) [MIM:300376]. BMD resembles DMD in hereditary and clinical features but is later in onset and more benign.
Defects in DMD are a cause of cardiomyopathy dilated X-linked type 3B (CMD3B) [MIM:302045]; also known as X-linked dilated cardiomyopathy (XLCM). Dilated cardiomyopathy is a disorder characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia. Patients are at risk of premature death.

Sequence similarities
Contains 2 CH (calponin-homology) domains.
Contains 22 spectrin repeats.
Contains 1 WW domain.
Contains 1 ZZ-type zinc finger.

Cellular localization
Cell membrane > sarcolemma. Cytoplasm > cytoskeleton.
Anti-Dystrophin antibody [MANDRA1] (ab7164) at 1/300 dilution +
whole lysate prepared from mouse myotubes at 30 µg

Secondary
Rabbit Anti-Mouse IgG H&L (HRP) (ab6728) at 1/8000 dilution

Developed using the ECL technique.

Exposure time: 5 minutes

This image was generated using the ascites version of the product.

ab7164 staining Dystrophin in frozen human tongue tissue sections
by Immunohistochemistry (IHC - Fr- Frozen sections). Samples
were incubated 1:100 dilution. A Goat Anti-mouse, FITC- conjugate
was used as the secondary antibody.

This image was generated using the ascites version of the product.

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