

Product datasheet

Anti-Dystrophin antibody [1808], prediluted ab75122

1 References

Overview

Product name	Anti-Dystrophin antibody [1808], prediluted
Description	Mouse monoclonal [1808] to Dystrophin, prediluted
Host species	Mouse
Specificity	ab75122 is highly specific to Dystrophin and shows no cross reaction with C protein (an isoform of alpha actinin), alpha actin, or human muscle spectrin.
Tested applications	Suitable for: IHC-P
Species reactivity	Reacts with: Mouse, Rat, Chicken, Human, Xenopus laevis, Torpedo
Immunogen	Acetylcholine receptor (AChR) enriched membranes and peripheral membrane proteins from Torpedo nobiliana electric organ.
Positive control	Skeletal muscle tissue.

Properties

Form	Prediluted
Storage instructions	Shipped at 4°C. Store at +4°C.
Storage buffer	Preservative: 15mM Sodium Azide Constituents: 0.5M Tris HCl, stabilizing protein, pH 7.6
Clonality	Monoclonal
Clone number	1808
Isotype	IgG1

Applications

Our [Abpromise guarantee](#) covers the use of **ab75122** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
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IHC-P

Application notes	IHC-P: Ready to use.
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Staining of formalin fixed tissues requires boiling tissue sections in 1mM EDTA, pH 8.0, for 10-20 minutes followed by cooling at room temperature for 20 minutes.

Not yet tested in other applications.

Optimal dilutions/concentrations should be determined by the end user.

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.

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