abcam

Product datasheet

Anti-Dystrophin antibody [1808], prediluted ab75122

1 References

概述

产品名称	Anti-Dystrophin抗体[1808], prediluted
描述	小鼠单克隆抗体[1808] to Dystrophin, prediluted
宿主	Mouse
特异性	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.
经测试应用	**适用于:** IHC-P
种属反应性	**与反应:** Mouse, Rat, Chicken, Human, Xenopus laevis, Torpedo
免疫原	Acetylcholine receptor (AChR) enriched membranes and peripheral membrane proteins from Torpedo nobiliana electric organ.
阳性对照	Skeletal muscle tissue.

性能

形式	Prediluted
存放说明	Shipped at 4°C. Store at +4°C.
存储溶液	Preservative: 15mM Sodium Azide Constituents: 0.5M Tris HCl, stabilizing protein, pH 7.6
克隆	单克隆
克隆编号	1808
同种型	IgG1

应用

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.

应用	Ab评论	说明
IHC-P		

应用说明	IHC-P: Ready to use.

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.

靶标

功能	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.
组织特异性	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.
疾病相关	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.
序列相似性	Contains 2 CH (calponin-homology) domains. Contains 22 spectrin repeats. Contains 1 WW domain. Contains 1 ZZ-type zinc finger.
细胞定位	Cell membrane > sarcolemma. Cytoplasm > cytoskeleton.

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