

## 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
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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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