Dystrophin (PT0821R) PT™ Rabbit mAb

CatalogNo: YM8580 Recombinant

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse,Rat

Applications
- WB,IHC,IF,IP,ELISA

MW
- 427kD (Calculated)

427kD (Observed)

Isotype
- IgG,Kappa

Recommended Dilution Ratios

IHC 1:200-1:1000
WB 1:500-1:2000
IF 1:200-1:1000
ELISA 1:5000-1:20000
IP 1:50-1:200

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA

Basic Information

Clonality	Monoclonal
Clone Number	PT0821R

Immunogen Information

Specificity	Endogenous

Target Information

Gene name DMD

Protein Name Dystrophin

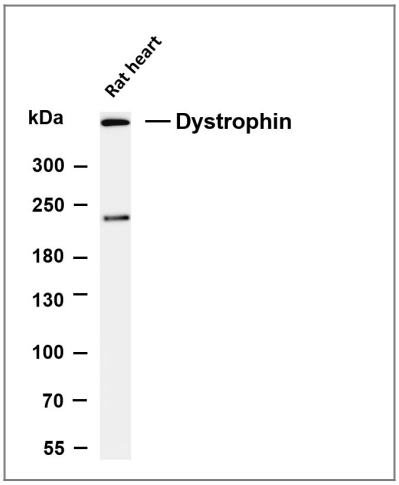
Organism	Gene ID	UniProt ID
Human	1756;	P11532;
Mouse		P11531;
Rat		P11530;

Cellular Localization Cell membrane, sarcolemma ; Peripheral membrane protein ; Cytoplasmic side . Cytoplasm, cytoskeleton . Cell junction, synapse, postsynaptic cell membrane . In muscle cells, sarcolemma localization requires the presence of ANK2, while localization to costameres requires the presence of ANK3. Localizes to neuromuscular junctions (NMJs). In adult muscle, NMJ localization depends upon ANK2 presence, but not in newborn animals. .

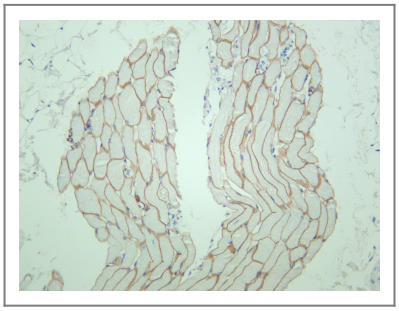
Tissue specificity Expressed in muscle fibers accumulating in the costameres of myoplasm at the sarcolemma. Expressed in brain, muscle, kidney, lung and testis. Most tissues contain transcripts of multiple isoforms. Isoform 15: Only isoform to be detected in heart and liver and is also expressed in brain, testis and hepatoma cells.

Function Alternative products:Additional isoforms seem to exist,Disease: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.,Disease: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.,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.,Function:May play a role in anchoring the cytoskeleton to the plasma membrane.,miscellaneous:The DMD gene is the largest known gene in humans. It is 2.4 million base-pairs in size, comprises 79 exons and takes over 16 hours to be transcribed and cotranscriptionally spliced.,online information:Dystrophin entry,online information:Dystrophin Mutation Database,online information:The Singapore human mutation and polymorphism database,similarity:Contains 1 WW domain.,similarity:Contains 1 ZZ-type zinc finger.,similarity:Contains 2 CH (calponin-homology) domains.,similarity:Contains 22 spectrin repeats.,subunit:Interacts with the syntrophins SNTA1, SNTB1, SNTB2, SNTG1 and SNTG2. Interacts with KRT19. Interacts with SYNM.,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.,

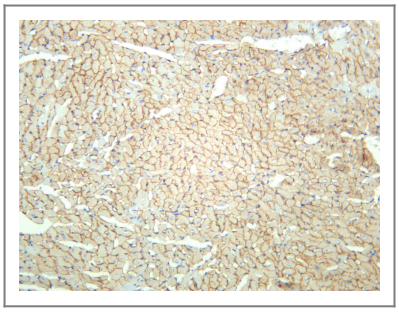
Validation Data



Various whole cell lysates were separated by 4-8% SDS-PAGE, and the membrane was blotted with anti-Dystrophin antibody. The HRP-conjugated Goat anti-Rabbit IgG (H + L) antibody was used to detect the antibody. Lane 1: Rat heart Predicted band size: 427kDa Observed band size: 427kDa



Human skeletal muscle was stained with anti-Dystrophin rabbit antibody



Mouse cardiac muscle was stained with anti-Dystrophin rabbit antibody

Contact information

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Please scan the QR code to access additional product information:
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