

Anti-Dystrophin Antibody [PSH26-84] - BSA and Azide free (Detector)

HA724649



Product Type:	Recombinant Rabbit monoclonal IgG, primary antibodies
Species reactivity:	Human, Mouse, Rat
Applications:	ELISA(Det)
Clone number:	PSH26-84

Description: Dystrophin is a rod-shaped cytoplasmic protein, and a vital part of a protein complex that connects the cytoskeleton of a muscle fiber to the surrounding extracellular matrix through the cell membrane. This complex is variously known as the costamere or the dystrophin-associated protein complex (DAPC). Many muscle proteins, such as α -dystrobrevin, syncoilin, synemin, sarcoglycan, dystroglycan, and sarcospan, colocalize with dystrophin at the costamere. It has a molecular weight of 427 kDa. Dystrophin is coded for by the DMD gene – the largest known human gene, covering 2.4 megabases (0.08% of the human genome) at locus Xp21. The primary transcript in muscle measures about 2,100 kilobases and takes 16 hours to transcribe; the mature mRNA measures 14.0 kilobases. The 79-exon muscle transcript codes for a protein of 3685 amino acid residues. Spontaneous or inherited mutations in the dystrophin gene can cause different forms of muscular dystrophy, a disease characterized by progressive muscular wasting. The most common of these disorders caused by genetic defects in dystrophin is Duchenne muscular dystrophy.

Immunogen: Recombinant protein within Human Dystrophin aa 3,365-3,685 (HA211485).

Positive control: Recombinant Human Dystrophin protein (HA211485).

Subcellular location: Cell membrane, sarcolemma, Cytoplasm, cytoskeleton, Postsynaptic cell membrane.

Database links: SwissProt: P11532 Human | P11531 Mouse | P11530 Rat

Recommended Dilutions:

ELISA(Det) Use at an assay dependent concentration. Can be paired for Sandwich ELISA with Rabbit monoclonal [PSH26-83] to Dystrophin (Capture) (HA724648) and Recombinant Human Dystrophin protein (HA211485) as the standard. The reference range value is 19.5-5,000 pg/mL.

Storage Buffer: 1*PBS (pH7.4).

Storage Instruction: Store at 2-8°C. Avoid freeze.

Purity: Protein A affinity purified.

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Orders:0086-571-88062880

Technical:0086-571-89986345

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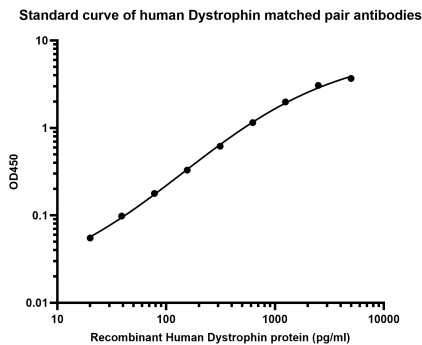
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Applications:WB=Western blot IHC-P=Immunohistochemistry (paraffin) IF-Cell=Immunofluorescence (Cell) IF-Tissue=Immunofluorescence (Tissue) FC=Flow cytometry IP=Immunoprecipitation

Images

Fig1: Sandwich ELISA analysis of Dystrophin matched pair antibodies

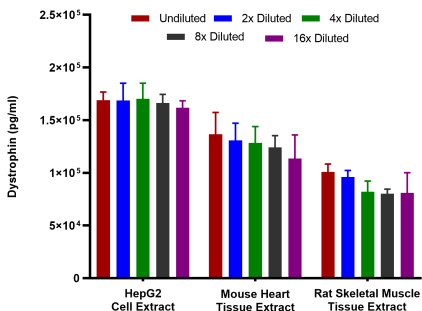
Capture: HA724648, Dystrophin Rabbit mAb [PSH26-83]
Detector: HA724649, Dystrophin Rabbit mAb [PSH26-84]



Elisa assay was performed by coating wells of a 96-well plate with 100 μ l per well of capture antibody (HA724648) diluted in carbonate/bicarbonate buffer, at a concentration of 2 μ g/mL overnight at 4 $^{\circ}$ C. Wells of the plate were washed, blocked with 150 μ l 0.05% tween-20 1% BSA blocking buffer, and incubated with serial diluted Recombinant Human Dystrophin protein (HA211485) starting from 5,000 pg/mL to 0 pg/mL and detect antibody (HA724649, Biotin, 0.2 μ g/mL) for 1 hour at 30 $^{\circ}$ C with shaking. Then the plate was washed and incubated with 100 μ l per well of SA-HRP for 0.5 hour at 30 $^{\circ}$ C with shaking. Detection was performed using an Ultra TMB Substrate for 10 minutes at room temperature in the dark. The reaction was stopped with sulfuric acid and absorbances were read on a spectrophotometer at 450 nm.

Fig2: Interpolated concentrations of native Dystrophin in HepG2, mouse heart and rat skeletal muscle extract samples based on a 1,000 μ g/mL extract load.

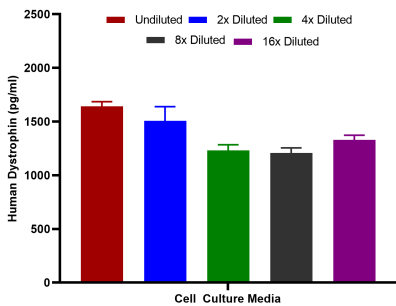
Capture: HA724648, Dystrophin Rabbit mAb [PSH26-83]
Detector: HA724649, Dystrophin Rabbit mAb [PSH26-84]



Interpolated concentration of native Dystrophin was measured in duplicate at different sample concentrations and interpolated from the Dystrophin standard curves. The interpolated dilution factor corrected values were plotted (mean $\pm$ SD, n=2). The mean Dystrophin concentration was determined to be 33,432 pg/mL in HepG2 cell extract, 12,667 pg/mL in mouse heart tissue extract and 8,798 pg/mL in rat skeletal muscle tissue extract.

Fig3: Interpolated concentrations of spiked Dystrophin in cell culture media samples.

Capture: HA724648, Dystrophin Rabbit mAb [PSH26-83]
Detector: HA724649, Dystrophin Rabbit mAb [PSH26-84]



The concentrations of Dystrophin were measured in duplicates, interpolated from the Dystrophin standard curves and corrected for sample dilution. Diluted samples are as follows: 50% cell culture media with FBS. The interpolated dilution factor corrected values are plotted (mean $\pm$ SD, n=2).

Note: All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE".

Background References

1. Fortunato F et al. The DMD gene and therapeutic approaches to restore dystrophin. Neuromuscul Disord. 2021 Oct
2. Bönnemann CG et al. Dystrophin Immunity after Gene Therapy for Duchenne's Muscular Dystrophy. N Engl J Med. 2023 Jun

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