

伴肌动蛋白抗体

产品货号： mIR2401

英文名称： Nebulin

中文名称： 伴肌动蛋白抗体

别名： DKFZp686C1456; FLJ11505; FLJ36536; FLJ39568; FLJ39584; NEB 177D; NEB; NEB177D; NEM 2; NEM2; NEBU_HUMAN; FLJ11505.

研究领域： 细胞生物 免疫学 结合蛋白

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Chicken, Dog, Pig, Cow, Rabbit,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

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

分子量： 500-800kDa

细胞定位： 细胞浆

性状： Lyophilized or Liquid

浓度： 1mg/ml

免疫原： KLH conjugated synthetic peptide derived from human Nebulin:6401-6669/6669

亚型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍： Nebulin is a high molecular weight protein (500-800 kD), localized at the N2-line of skeletal muscle myofibrils. It exhibits an elastic stretch dependence, as it maintains the same proportional distance between the z-line and the m-line as the sarcomere is stretched.

Function:

This giant muscle protein may be involved in maintaining the structural integrity of sarcomeres and the membrane system associated with the myofibrils. Bind and stabilize F-actin.

Subcellular Location:

Cytoplasm > myofibril > sarcomere. Cytoplasm > cytoskeleton.

Tissue Specificity:

Muscle specific. Located in the thin filament of striated muscle.

DISEASE:

Defects in NEB are the cause of nemaline myopathy type 2 (NEM2) [MIM:256030]. A form of nemaline myopathy. Nemaline myopathies are muscular disorders characterized by muscle weakness of varying severity and onset, and abnormal thread-or rod-like structures in muscle fibers on histologic examination.

Similarity:

Contains 178 nebulin repeats.

Contains 1 SH3 domain.

SWISS:

P20929

Gene ID:

4703

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

具有稳定细肌丝的作用。