abcam

Product datasheet

Recombinant Human heavy chain Myosin/MYH3 protein ab114308

1 图像

描述

产品名称	重组人heavy chain Myosin/MYH3蛋白
表达系统	Wheat germ
Accession	P11055
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Human
序列	SSDTEMEVFGIAAPFLRKSEKERIEAQNQPFDAKTYCFVVDS KEEYAKGK IKSSQDGKVTVETEDNRTLVVKPEDVYAMNPPKFDRIEDMAM LTHLNEP
预测分子量	37 kDa including tags
氨基酸	2 to 100

技术指标

Our **Abpromise guarantee** covers the use of **ab114308** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	SDS-PAGE ELISA Western blot
形式	Liquid
补充说明	This product was previously labelled as heavy chain Myosin.

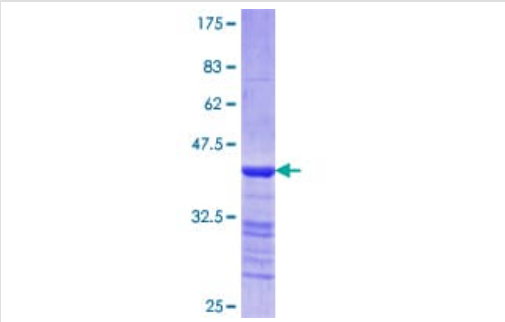
制备和贮存

稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

常规信息

功能	Muscle contraction.
疾病相关	Defects in MYH3 are the cause of distal arthrogryposis type 2A (DA2A) [MIM:193700]; also known as Freeman-Sheldon syndrome (FSS). Distal arthrogryposis is a clinically and genetically heterogeneous group of disorders characterized by bone anomalies and joint contractures of the hands and feet, causing medially overlapping fingers, clenched fists, ulnar deviation of fingers, camptodactyly and positional foot deformities. It is a disorder of primary limb malformation without primary neurologic or muscle disease. DA2A is the most severe form of distal arthrogryposis. Affected individuals have contractures of the orofacial muscles, characterized by microstomia with pouting lips, H-shaped dimpling of the chin, deep nasolabial folds, and blepharophimosis. Dysphagia, failure to thrive, growth deficit, and life-threatening respiratory complications (caused by structural anomalies of the oropharynx and upper airways) are frequent. Inheritance is autosomal dominant. Defects in MYH3 are the cause of distal arthrogryposis type 2B (DA2B) [MIM:601680]; also known as Sheldon-Hall syndrome (SHS) or arthrogryposis multiplex congenita distal type 2B (AMCD2B). DA2B is a form of inherited multiple congenital contractures. Affected individuals have vertical talus, ulnar deviation in the hands, severe camptodactyly, and a distinctive face characterized by a triangular shape, prominent nasolabial folds, small mouth and a prominent chin. DA2B is the most common of the distal arthrogryposis syndromes. It is similar to DA2A but the facial contractures are less dramatic.
序列相似性	Contains 1 IQ domain. Contains 1 myosin head-like domain.
发展阶段	Abundantly present in fetal skeletal muscle and not present or barely detectable in heart and adult skeletal muscle.
结构域	The rodlike tail sequence is highly repetitive, showing cycles of a 28-residue repeat pattern composed of 4 heptapeptides, characteristic for alpha-helical coiled coils. Each myosin heavy chain can be split into 1 light meromyosin (LMM) and 1 heavy meromyosin (HMM). It can later be split further into 2 globular subfragments (S1) and 1 rod-shaped subfragment (S2).
细胞定位	Cytoplasm > myofibril. Thick filaments of the myofibrils.

图片



SDS-PAGE - Recombinant Human heavy chain Myosin/MYH3 protein (ab114308)

SDS-PAGE analysis of ab114308 on a 12.5% gel stained with Coomassie Blue.

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