

## Product datasheet

# Recombinant Human heavy chain Myosin/MYH3 protein ab114308

画像数 1

### 製品の詳細

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製品名	Recombinant Human heavy chain Myosin/MYH3 protein	
発現系	Wheat germ	
アクセッション番号	<b>P11055</b>	
タンパク質長	Protein fragment	
Animal free	No	
由来	Recombinant	
生物種	Human	
配列	SSDTEMEVFGIAAPFLRKSEKERIEAQNQPFDKTYCFVVDS KEEYAKGK IKSSQDGKVTVETEDNRTLTVVKPEDVYAMNPPKFDRIEDMAM LTHLNEP	
予測される分子量	37 kDa including tags	
領域	2 to 100	

### 特性

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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.

アプリケーション	SDS-PAGE ELISA Western blot
製品の状態	Liquid
備考	This product was previously labelled as heavy chain Myosin.

### 前処理および保存

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保存方法および安定性	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl
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## 関連情報

### 機能

Muscle contraction.

### 関連疾患

Defects in MYH3 are the cause of distal arthrogyposis type 2A (DA2A) [MIM:193700]; also known as Freeman-Sheldon syndrome (FSS). Distal arthrogyposis 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 arthrogyposis. 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 arthrogyposis type 2B (DA2B) [MIM:601680]; also known as Sheldon-Hall syndrome (SHS) or arthrogyposis 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 arthrogyposis syndromes. It is similar to DA2A but the facial contractures are less dramatic.

### 配列類似性

Contains 1 IQ domain.

Contains 1 myosin head-like domain.

### 発生段階

Abundantly present in fetal skeletal muscle and not present or barely detectable in heart and adult skeletal muscle.

### ドメイン

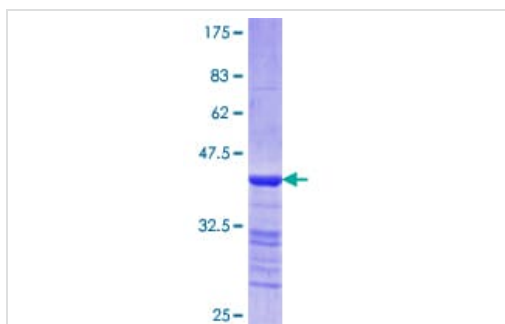
The rodlike tail sequence is highly repetitive, showing cycles of a 28-residue repeat pattern composed of 4 heptapeptides, characteristic for alpha-helical coiled coils.

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).

### 細胞内局在

Cytoplasm > myofibril. Thick filaments of the myofibrils.

## 画像



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

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

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

### **Our Abpromise to you: Quality guaranteed and expert technical support**

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- Response to your inquiry within 24 hours
  
- We provide support in Chinese, English, French, German, Japanese and Spanish
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