# abcam

# Product datasheet

# Recombinant Human heavy chain Myosin/MYH3 protein ab114308

# 1 Image

**Description** 

**Product name** Recombinant Human heavy chain Myosin/MYH3 protein

Expression system Wheat germ
Accession P11055

Protein length Protein fragment

Animal free No

**Nature** Recombinant

**Species** Human

Sequence SSDTEMEVFGIAAPFLRKSEKERIEAQNQPFDAKTYCFVV

DSKEEYAKGK

**IKSSQDGKVTVETEDNRTLVVKPEDVYAMNPPKFDRIEDM** 

AMLTHLNEP

Predicted molecular weight 37 kDa including tags

Amino acids 2 to 100

# **Specifications**

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.

**Applications** SDS-PAGE

**ELISA** 

Western blot

Form Liquid

Additional notes This product was previously labelled as heavy chain Myosin.

**Preparation and Storage** 

Stability and Storage 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 HCI

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#### **General Info**

#### **Function**

Muscle contraction.

#### Involvement in disease

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.

#### Sequence similarities

Contains 1 IQ domain.

Contains 1 myosin head-like domain.

#### **Developmental stage**

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

skeletal muscle.

# **Domain**

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

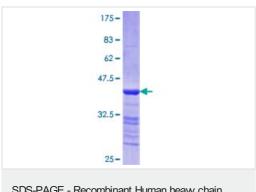
(HMM). It can later be split further into 2 globular subfragments (S1) and 1 rod-shaped

subfragment (S2).

#### **Cellular localization**

Cytoplasm > myofibril. Thick filaments of the myofibrils.

#### **Images**



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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