

Actin, sarcomeric muscle (ABT55R) rabbit mAb

Catalog No :	YM7003
Reactivity :	Human; Mouse (predicted: Rat; Bovin; Pig; Chick)
Applications :	WB; IHC; ELISA
Target :	Actin, sarcomeric muscle
Gene Name :	ACTA1;ACTA;ACTC1;ACTC
Protein Name :	Actin, sarcomeric muscle
Human Gene Id :	375/377
Human Swiss Prot No :	P68032
Immunogen :	Synthesized peptide derived from human Actin, sarcomeric muscle AA range:2-50
Specificity :	This antibody detects endogenous levels of Actin, sarcomeric muscle
Formulation :	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source :	Monoclonal, Rabbit IgG1, Kappa
Dilution :	IHC 1:100-500, WB 1:500-1000, ELISA 1:5000-20000
Purification :	Recombinant Expression and Affinity purified
Storage Stability :	-15°C to -25°C/1 year(Do not lower than -25°C)
Molecularweight :	42kDa
Background :	The product encoded by this gene belongs to the actin family of proteins, which are highly conserved proteins that play a role in cell motility, structure and integrity. Alpha, beta and gamma actin isoforms have been identified, with alpha actins being a major constituent of the contractile apparatus, while beta and gamma actins are involved in the regulation of cell motility. This actin is an alpha

actin that is found in skeletal muscle. Mutations in this gene cause nemaline myopathy type 3, congenital myopathy with excess of thin myofilaments, congenital myopathy with cores, and congenital myopathy with fiber-type disproportion, diseases that lead to muscle fiber defects. [provided by RefSeq, Jul 2008],

Function :

disease:Defects in ACTA1 are a cause of congenital myopathy with excess of thin myofilaments (CM) [MIM:102610].,disease:Defects in ACTA1 are a cause of congenital myopathy with fiber-type disproportion (CFTD) [MIM:255310]; also known as congenital fiber-type disproportion myopathy (CFTDM). CFTD is a genetically heterogeneous disorder in which there is relative hypotrophy of type 1 muscle fibers compared to type 2 fibers on skeletal muscle biopsy. However, these findings are not specific and can be found in many different myopathic and neuropathic conditions.,disease:Defects in ACTA1 are the cause of nemaline myopathy type 3 (NEM3) [MIM:161800]. Nemaline myopathy (NEM) is a form of congenital myopathy characterized by abnormal thread- or rod-like structures in muscle fibers on histologic examination. The clinical phenotype is highly variable, with differing age at onset and severity.,func

Subcellular Location :

Cytoplasm, cytoskeleton.

Expression :

Epithelium,Skeletal muscle,

Products Images