

Datasheet for ABIN2804352

anti-MTM1 antibody (AA 225-275) (AbBy Fluor® 594)



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Overview

Quantity:	100 µL
Target:	MTM1
Binding Specificity:	AA 225-275
Reactivity:	Human, Mouse, Rat
Host:	Rabbit
Clonality:	Polyclonal
Conjugate:	This MTM1 antibody is conjugated to AbBy Fluor® 594
Application:	Western Blotting (WB), Flow Cytometry (FACS), Immunofluorescence (Paraffin-embedded Sections) (IF (p))

Product Details

Immunogen:	KLH conjugated synthetic peptide derived from human MTM1/Myotubularin
Isotype:	IgG
Cross-Reactivity:	Human, Mouse, Rat
Purification:	Purified by Protein A.

Target Details

Target:	MTM1
Alternative Name:	MTM1 (MTM1 Products)
Background:	Synonyms: CG2, CNM, KIAA4176, mKIAA4176, Mtm, Mtm1, MTM1_HUMAN, MTMX,

Target Details

Myotubular myopathy 1, Myotubularin, XLMTM.

Background: X-linked recessive myotubular myopathy is a congenital muscular disease characterized by severe hypotonia and generalized muscle weakness that, in most cases, leads to early postnatal death. The gene responsible for myotubular myopathy MTM1 encodes a dual specificity phosphatase, named myotubularin, which is highly conserved through evolution. The gene for MTM1 is localized to a 300 kb critical region on human Xq128 between IDS and GRBRA3. Human MTM1, a 603 amino-acid protein, is mutated in myotubular myopathy. The largely related protein hMTMR2 is found mutated in a recessive form of Charcot-Marie-Tooth neuropathy. Myotubularin is primarily a lipid phosphatase that acts on phosphatidylinositol 3-monophosphate and is involved in the regulation of the phosphatidylinositol 3-kinase (PI3-kinase) pathway and membrane trafficking. Wild-type myotubularin can directly dephosphorylate PI3P and PI4P in vitro. Thus, it decreases PI3P levels by down-regulating PI3K activity and by facilitating the degradation of PI3P.

Gene ID: 4534

UniProt: [Q13496](#)

Pathways: [Inositol Metabolic Process](#), [Skeletal Muscle Fiber Development](#)

Application Details

Application Notes: FCM 1:20-100
IF(IHC-P) 1:50-200

Restrictions: For Research Use only

Handling

Format: Liquid

Concentration: 1 µg/µL

Buffer: Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol.

Preservative: ProClin

Precaution of Use: This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only.

Storage: -20 °C

Handling

Storage Comment: Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.

Expiry Date: 12 months