

bs-9178R**[Primary Antibody]****MTM1 Rabbit pAb****BioSS**
ANTIBODIES

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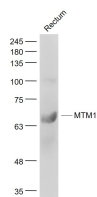
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— DATASHEET —

Host: Rabbit	Isotype: IgG	Applications: WB (1:500-2000)
Clonality: Polyclonal		Reactivity: Mouse (predicted: Human, Rat, Rabbit, Pig, Dog)
GeneID: 4534	SWISS: Q13496	
Target: MTM1		Predicted MW.: 70 kDa
Immunogen: KLH conjugated synthetic peptide derived from human MTM1/Myotubularin: 201-300/603.		Subcellular Location: Cell membrane ,Cytoplasm
Purification: affinity purified by Protein A		
Concentration: 1mg/ml		
Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4°C. Store at -20°C for one year. Avoid repeated freeze/thaw cycles.		
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.		

— VALIDATION IMAGES —

Sample: Rectum (Mouse) Lysate at 40 ug
Primary: Anti- MTM1 (bs-9178R) at 1/1000
dilution Secondary: IRDye800CW Goat Anti-
Rabbit IgG at 1/20000 dilution Predicted band
size: 70 kD Observed band size: 68 kD