CLCN1 Monoclonal Antibody

Catalog No	BYmab-05462
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB
Gene Name	CLCN1 CLC1
Protein Name	Chloride channel protein 1 (ClC-1) (Chloride channel protein, skeletal muscle)
Immunogen	Synthesized peptide derived from part region of human protein
Specificity	CLCN1 Monoclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	108kD
Cell Pathway	Cell membrane ; Multi-pass membrane protein .
Tissue Specificity	Predominantly expressed in skeletal muscles.
Function	disease:Defects in CLCN1 are the cause of autosomal dominant myotonia congenita (MCD) [MIM:160800]; also known as Thomsen disease. MCD is characterized by skeletal muscle stiffness (delayed relaxation), due to membrane hyperexcitability. A variant form of Thomsen disease is myotonia levior that is characterized by milder symptoms, later onset and absence of muscle hypo- and hypertrophy.,disease:Defects in CLCN1 are the cause of autosomal recessive myotonia congenita (MCR) [MIM:255700]; also known as Becker disease.,function:Voltage-gated chloride channel. Chloride channels have several functions including the regulation of cell volume; membrane potential stabilization, signal transduction and transepithelial transport.,miscellaneous:The CLC channel family contains both chloride channels and proton-coupled anion transporters that exchange chloride or another anion for protons. The absence

Background	chloride voltage-gated channel 1(CLCN1) Homo sapiens The CLCN family of voltage-dependent chloride channel genes comprises nine members (CLCN1-7, Ka and Kb) which demonstrate quite diverse functional characteristics while sharing significant sequence homology. The protein encoded by this gene regulates the electric excitability of the skeletal muscle membrane. Mutations in this gene cause two forms of inherited human muscle disorders: recessive generalized myotonia congenita (Becker) and dominant myotonia (Thomsen). Alternative splicing results in multiple transcript variants. [provided by RefSeq, Mar 2012],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

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