10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

hCLC1-G972 Blocking Peptide

Synthetic peptide
Catalog # BP6329c

Specification

hCLC1-G972 Blocking Peptide - Product Information

Primary Accession	P35523
Other Accession	P35524, Q64347

hCLC1-G972 Blocking Peptide - Additional Information

Gene ID 1180

Other Names
Chloride channel protein 1, ClC-1, Chloride channel protein, skeletal muscle, CLCN1, CLC1

Target/Specificity
The synthetic peptide sequence is selected from aa 972~988 of HUMAN CLCN1

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

hCLC1-G972 Blocking Peptide - Protein Information

Name CLCN1 {ECO:0000303|PubMed:8533761, ECO:0000312|HGNC:HGNC:2019}

Function
Voltage-gated chloride channel involved in skeletal muscle excitability. Generates most of the plasma membrane chloride conductance in skeletal muscle fibers, stabilizes the resting membrane potential and contributes to the repolarization phase during action potential firing (PubMed:12456816, PubMed:16027167, PubMed:22521272, PubMed:22641783, PubMed:26007199, PubMed:26502825, PubMed:26510092, PubMed:7951242, PubMed:8112288, PubMed:8130334, PubMed:9122265, PubMed:<a

href="http://www.uniprot.org/citations/9565403" target="_blank">9565403, PubMed:9736777). Forms a homodimeric channel where each subunit has its own ion conduction pathway. Conducts double-barreled currents controlled by two types of gates, two fast glutamate gates that control each subunit independently and a slow common gate that opens and shuts off both subunits simultaneously. Has a significant open probability at muscle resting potential and is further activated upon membrane depolarization (PubMed:10051520, PubMed:10962018, PubMed:29809153, PubMed:31022181). Permeable to small monovalent anions with ion selectivity for chloride > thiocyanate > bromide > nitrate > iodide (PubMed:9122265, PubMed:9565403).

Cellular Location
Cell membrane; Multi-pass membrane protein Cell membrane, sarcolemma {ECO:0000250|UniProtKB:Q64347}; Multi-pass membrane protein. Cell membrane, sarcolemma, T-tubule {ECO:0000250|UniProtKB:Q64347}; Multi-pass membrane protein

Tissue Location
Predominantly expressed in skeletal muscles.

hCLC1-G972 Blocking Peptide - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Blocking Peptides

hCLC1-G972 Blocking Peptide - Images

hCLC1-G972 Blocking Peptide - Background

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).

hCLC1-G972 Blocking Peptide - References

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Hebeisen, S., et al., J. Biol. Chem. 279(13):13140-13147 (2004).
Letizia, C., et al., Calcif. Tissue Int. 74(1):42-46 (2004).
Estevez, R., et al., Neuron 38(1):47-59 (2003).
Pusch, M., Hum. Mutat. 19(4):423-434 (2002).