ALPHA DIAGNOSTIC INTERNATIONAL

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Product Specification Sheet

Chloride Channle-1 (CLC-1 or CLCN1) Antibodies

Cat. # CLC11-PRat CLC1 Control Peptide #1	**SIZE**: 100 ug
Cat. # CLC11-SRabbit Anti-rat CLC-1 antiserum #1	**SIZE**: 100 ul
Cat. # CLC11-ARabbit Anti-rat CLC-1 IgG #1 (aff pure)	**SIZE**: 100 ug

Chloride is a critical component of all living cells. Voltage-gated chloride channels regulate cellular traffic of chloride ion. The chloride channels (CIC or CLC) performs several functions including the regulation of cell volume, membrane potential stabilization, signal transduction, and transepithelial transport. Mutations in CIC genes have been linked with several human diseases including myotonias (Thomsen's disease), cystic fibrosis, Bartters syndrome type III, Dent's disease, and X-linked recessive nephrolithiasis. In mammals, CLC proteins form a superfamily of at least 9 different genes (CLC1-7 also known as CLCN1-7 and CLK1-2 or CLCKa and CLCKb). Additional forms of these proteins are obtained by alternative splicing. All CLC proteins (~700-1000 aa) are predicted to contain 10 (possibly 12) transmembrane domains. Except CLC-1 and CLC-K1/K2 that are specific for kidney, most other CLC are widely distributed in various tissues.

Rat CLC-1 is 994 aa membrane protein (human CLC-1 988 aa) (1). It is predominantly expressed in skeletal muscles. Defects in CLC1 (CLCN1) are the cause of autosomal recessive generalized myotonia (Becker's disease) (RGM) and autosomal dominant myotonia congenita (Thomsen's disease; MC) which are characterized by skeletal muscle stiffness (delayed relaxation) that is a result of muscle membrane hyperexcitability

Source of Antigen and Antibodies

Antigen	18-aa peptide of rat CLC1 A; **Designated (CLC11-P or control peptide/blocking peptide**) conjugated to KLH; epitope location ~C-terminus, extracellular domain
Ab Host/type	Rabbit, Polyclonal unpurified antiserum (#**CLC11**-S) and IgG, purified over antigen-agarose (Cat # **CLC11**-A)
2-Ab	Cat # 20320, goat anti-rabbit IgG-HRP (AP, biotin, FITC conjugates also available).
-ve control IgG	# 20009-1, Rabbit (non-immune) IgG, purified, suitable for ELISA, Western, IHC as –ve control

Form & Storage of Antibodies/Peptide Control

Antiserum (unpurified)
100ul solution lyophilized powder
Supplied 0.05% azide, **Reconstitute** powder in 100 ul PBS

Affinity pure IgG
100 ug/100ul solution lyophilized powder
Supplied in **Buffer**: PBS+0.1% BSA
Reconstitute powder in PBS at 1mg/ml

Control/blocking peptide
100 ug/100 ul solution lyophilized powder
Supplied in Buffer: PBS pH 7.5,
Reconstitute powder in PBS at 1 mg/ml.

Storage
Short-term: unopened, undiluted liquid vials at -20OC and powder at 4oC or -20oC..
Long-term: at –20C or below in suitable aliquots after reconstitution. Do not freeze and thaw and store working, diluted solutions.

Stability: 6-12 months at –20oC or below.
Shipping: 4oC for solutions and room temp for powder

Recommended Usage

Western Blotting (1:1K-5K for neat serum and 1-10 ug/ml for affinity pure antibody using ECL technique). See refs in 2

ELISA: Control peptide can be used to coat ELISA plates at 1 ug/ml and detected with antibodies (1:10-50K for neat serum and 0.5-1 ug/ml for affinity pure).

Histochemistry & Immunofluorescence: We recommend the use of affinity purified antibody at 1-20 ug/ml in paraformaldehyde fixed sections of tissues. See refs in 2

Specificity & Cross-reactivity

The 18 AA rat CLC11-P control peptide 100% conserved in mouse, dog, rat, and human CLC-1. No significant sequence homology is detected with other CLCs or other proteins. Antibody cross-reactivity in various species has not been studied. Control peptide, because of its low mol. Wt (<3 kDa), is not suitable for Western. It should be used for ELISA or antibody blocking experiments (use 5-10 ug control peptide per 1 ug of aff pure IgG or 1 ul antiserum) to confirm antibody specificity (see detailed protocol at the web site).

General References: Steinmeyer K (1991) Nature 354, 301; Koch MC (1992) Science 257, 797; George AL (1993) Nature Genet. 3, 305; Steinmeyer K (1994) EMBO J 13, 737; Lorenz C (1994) Human Mol. Genet. 3, 941; Heine R (1994) Hum. Mol. Genet. 3, 1123; George Al (1994) Hum. Mol. Genet. 3, 2071; Meyer-Kleine C (1995) Am. J. Hum. Genet. 57, 1325; Lehman Horn F (1995) Hum. Mol. Genet 4, 1397

2. Citations for ADI Antibodies (see updates at the web site)

Wheeler TM, 2007, J. Clin. Invest., 117: 3952 - 3957. IF
Kanadia RN, 2006, PNAS, 103: 11748 - 11753 WB IHC
Yu Z, 2006, J. Clin. Invest., Oct 2006; 116: 2663 – 2672, IF

*This product is for In vitro research use only.

Related material available from ADI

Antibodies CLC1-7/CLCK1, KCC1-3; AQP-9 and RUT; OCT and OAT, AE-3, and NACX

CLC11-S-A-P 71217A

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