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CLCNKB Rabbit pAb

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Catalog # AP55225

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P51801
Predicted	Rat, Pig, Dog, Human, Mouse, Rabbit
Host	Rabbit
Clonality	Polyclonal
Calculated MW	75446
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CLCNKB
Epitope Specificity	51-150/687
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cell membrane.
DISEASE	Defects in CLCNKB are the cause of Bartter syndrome type 3 (BS3) [MIM:607364]; also known as classic Bartter syndrome. It is an autosomal recessive form of often severe intravascular volume depletion due to renal salt-wasting associated with low blood pressure, hypokalemic alkalosis, hypercalciuria, and normal serum magnesium levels. Defects in CLCNKB are a cause of Bartter syndrome type 4B (BS4B) [MIM:613090]. A digenic, recessive disorder characterized by impaired salt reabsorption in the thick ascending loop of Henle with pronounced salt wasting, hypokalemic metabolic alkalosis, and varying degrees of hypercalciuria. Bartter syndrome type 4B is associated with sensorineural deafness.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The family of voltage-dependent chloride channels (CLCs) regulate cellular trafficking of chloride ions, a critical component of all living cells. CLCs regulate excitability in muscle and nerve cells, aid in organic solute transport, and maintain cellular volume. CLC-KA is a kidney-specific chloride channel that mediates transepithelial chloride transport in the thin ascending limb of the Henle loop in the inner medulla. CLC-KA plays a crucial role in urine concentration. The gene encoding human CLC-KA maps to chromosome 1p36. Mutations in this gene may be associated with nephrogenic diabetes insipidus in those cases where mutations in the vasopressin V2 receptor and the AQP2 water channel are lacking. CLC-KB mediates basolateral chloride ion efflux in the thick ascending limb and in more distal nephron segments. The gene encoding human CLC-KB maps to chromosome 1p36. Mutations in this gene cause type III Barter's syndrome which is characterized by renal salt-wasting and low blood pressure.

Additional Information

Gene ID	1188
Other Names	Chloride channel protein ClC-Kb, Chloride channel Kb, ClC-K2, CLCNKB {ECO:0000303|PubMed:18310267, ECO:0000312|HGNC:HGNC:2027}
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	CLCNKB {ECO:0000303|PubMed:18310267, ECO:0000312|HGNC:HGNC:2027}
Function	Anion-selective channel permeable to small monovalent anions with ion selectivity for chloride > bromide > nitrate > iodide (PubMed:11734858, PubMed:12111250). Forms a homodimeric channel where each subunit has its own ion conduction pathway. May conduct double- barreled currents controlled by two types of gates, two fast gates that control each subunit independently and a slow common gate that opens and shuts off both subunits simultaneously (PubMed:11734858, PubMed:12111250, PubMed:16849430, PubMed:18776122, PubMed:19646679). Assembles with the regulatory subunit BSND/Barttin for sorting at the basolateral plasma membrane domain and functional switch to the ion conducting state. CLCNKB:BSND channels display mostly a linear current- voltage relationship controlled by common gate (PubMed:11734858, PubMed:12111250, PubMed:16849430, PubMed:18776122, PubMed:19646679). Mediates chloride conductance along nephron segments, namely the thick ascending limb of Henle's loop, convoluted tubule and the collecting duct, contributing to the maintenance of systemic acid-base and electrolyte homeostasis (By similarity). Conducts chloride currents in the stria vascularis of the inner ear to establish the endocochlear potential necessary for normal hearing (PubMed:15044642, PubMed:18310267, PubMed:19646679).
Cellular Location	Basolateral cell membrane; Multi-pass membrane protein

Background

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