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Rabbit Anti-CLCNKB antibody

SL13627R

Product Name:	CLCNKB
Chinese Name:	氯离子通道KB抗体
Alias:	Bartter syndrome type 3; Chloride channel Kb; Chloride channel kidney B; Chloride channel protein ClC-Kb; Chloride channel voltage sensitive Kb; ClC K2; ClC-K2; ClCK2; CLCKB; CLCKB_HUMAN; CLCNKB; hClC Kb; hClCKb; MGC24087; OTTHUMP00000011120; OTTHUMP00000011121; RP11 5P18.8.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	75kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CLCNKB:51-150/687
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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.

Function:
Voltage-gated chloride channel. Chloride channels have several functions including the regulation of cell volume; membrane potential stabilization, signal transduction and transepithelial transport. May be important in urinary concentrating mechanisms.

Subcellular Location:
Cell membrane.

Tissue Specificity:
Expressed predominantly in the kidney.

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.

Similarity:
Belongs to the chloride channel (TC 2.A.49) family. CLCNKB subfamily.
Contains 2 CBS domains.

SWISS:
P51801

Gene ID:
1188

Database links:

Entrez Gene: 1188 Human

	Entrez Gene: 56365 Mouse Entrez Gene: 79430 Rat Omim: 602023 Human SwissProt: P51801 Human SwissProt: Q9WUB6 Mouse SwissProt: P51802 Rat Unigene: 352243 Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.