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Rabbit Anti-Chloride Channel 5 antibody

SL10307R

Product Name:	Chloride Channel 5
Chinese Name:	氯离子Channel protein5抗体
Alias:	Chloride channel protein 5; Chloride transporter ClC-5; ClC-5; CLC5; CLCK2; CLCN5; CLCN5_HUMAN; DENTS; H(+)/Cl(-) exchange transporter 5; hCIC-K2; NPHL1; NPHL2; XLRH; XRN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Rabbit,Sheep,
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:	83kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Chloride Channel 5/CLC5:1-100/746
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. The genes encoding human CLC-1 through CLC-7 map to chromosomes 7q32, 3q28, 4q32,

Xp22.3, Xp11.23-p11.22, 1p36 and 16p13, respectively. CLC1 is highly expressed in skeletal muscle. Mutations in the gene encoding CLC1 lead to myotonia, an inheritable disorder characterized by muscle stiffness and renal salt wasting. CLC2 is highly expressed in the epithelia of several organs including lung, which suggests CLC2 may be a possible therapeutic target for cystic fibrosis. CLC3 expression is particularly abundant in neuronal tissue, while CLC4 expression is evident in skeletal and cardiac muscle as well as brain. Mutations in the gene encoding CLC5 lead to Dent's disease, a renal disorder characterized by proteinuria and hypercalciuria. CLC6 and CLC7 are broadly expressed in several tissues including testis, kidney, brain and muscle.

Function:
Proton-coupled chloride transporter. Functions as antiport system and exchanges chloride ions against protons. Important for normal acidification of the endosome lumen. May play an important role in renal tubular function.

Subcellular Location:
Golgi apparatus membrane. Endosome membrane. Cell membrane.

Tissue Specificity:
Kidney. Moderately expressed in aortic vascular smooth muscle and endothelial cells, and at a slightly higher level in the coronary vascular smooth muscle.

Post-translational modifications:
Ubiquitinated by NEDD4L in the presence of albumin; which promotes endocytosis and proteasomal degradation.

DISEASE:
Defects in CLCN5 are a cause of hypophosphatemic rickets, X-linked recessive (XLRHR) [MIM:300554]. XLRHR is a renal disease belonging to the 'Dent disease complex', a group of disorders characterized by proximal renal tubular defect, hypercalciuria, nephrocalcinosis, and renal insufficiency. The spectrum of phenotypic features is remarkably similar in the various disorders, except for differences in the severity of bone deformities and renal impairment. XLRH patients present with rickets or osteomalacia, hypophosphatemia due to decreased renal tubular phosphate reabsorption, hypercalciuria, and low molecular weight proteinuria. Patients develop nephrocalcinosis with progressive renal failure in adulthood. Female carriers may have asymptomatic hypercalciuria or hypophosphatemia only.
Defects in CLCN5 are the cause of nephrolithiasis type 2 (NPHL2) [MIM:300009]; also known as Dent disease 1. NPHL2 is an X-linked recessive renal disease belonging to the 'Dent disease complex'. NPHL2 patients manifest hypercalciuria, hypophosphatemia, aminoaciduria, nephrocalcinosis and nephrolithiasis, renal insufficiency leading to renal failure in adulthood, rickets (33% of patients) and osteomalacia.
Defects in CLCN5 are the cause of nephrolithiasis type 1 (NPHL1) [MIM:310468]; also designated XRN. NPHL1 is an X-linked recessive renal disease belonging to the 'Dent disease complex'. NPHL1 presents with hypercalciuria, nephrocalcinosis, renal stones and renal insufficiency. Patients lack urinary acidification defects, rickets, and

osteomalacia. Defects in CLCN5 are the cause of low molecular weight proteinuria with hypercalciuria and nephrocalcinosis (LMWPHN) [MIM:308990]. LMWPHN is an X-linked renal disease belonging to the 'Dent disease complex'. Patients tend to have hypercalciuric nephrocalcinosis without rickets or renal failure.

Similarity:
Belongs to the chloride channel (TC 2.A.49) family. ClC-5/CLCN5 subfamily. Contains 2 CBS domains.

SWISS:
P51795

Gene ID:
1184

Database links:

Entrez Gene: 1184Human

Entrez Gene: 12728Mouse

Entrez Gene: 25749Rat

Omim: 300008Human

SwissProt: P51795Human

SwissProt: Q9WVD4Mouse

SwissProt: P51796Rat

Unigene: 166486Human

Unigene: 745501Human

Unigene: 486257Mouse

Unigene: 10337Rat

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.