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[Primary Antibody]

Rabbit Anti-CLCN2 antibody

Catalog Number: SL6470R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	CLCN2
Immunogen:	KLH conjugated synthetic peptide derived from human CLCN2/CLC-2: 501-600/898
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB=1:500-2000 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Human(predicted:Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Rabbit)
Predicted MW.:	99kDa
Subcellular Location:	The cell membrane
Background:	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

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	characterized by proteinuria and hypercalciuria. CLC6 and CLC7 are broadly expressed in several tissues including testis, kidney, brain and muscle. **Function:** Voltage-gated chloride channel. Chloride channels have several functions including the regulation of cell volume; membrane potential stabilization, signal transduction and transepithelial transport. **Subcellular Location:** Membrane; Multi-pass membrane protein. **Tissue Specificity:** Ubiquitously expressed. Moderately expressed in aortic and coronary vascular smooth muscle cells and expressed at a low level in aortic endothelial cells. DISEASE: Defects in CLCN2 are associated with susceptibility to epilepsy, idiopathic generalized type 11 (EIG11) [MIM:607628]. A disorder characterized by recurring generalized seizures in the absence of detectable brain lesions and/or metabolic abnormalities. Generalized seizures arise diffusely and simultaneously from both hemispheres of the brain. Defects in CLCN2 are associated with juvenile absence epilepsy type 2 (JAE2) [MIM:607628]. JAE is a subtype of idiopathic generalized epilepsy (IGE) characterized by onset occurring around puberty, absence seizures, generalized tonic-clonic seizures (GTCS), GTCS on awakening and myoclonic seizures. Defects in CLCN2 are associated with juvenile myoclonic epilepsy type 8 (EJM8) [MIM:607628]. A subtype of idiopathic generalized epilepsy. Patients have afebrile seizures only, with onset in adolescence (rather than in childhood) and myoclonic jerks which usually occur after awakening and are triggered by sleep deprivation and fatigue. **Similarity:** Belongs to the chloride channel (TC 2.A.49) family. ClC-2/CLCN2 subfamily. Contains 2 CBS domains. **Database links:** **Entrez Gene:** 1181 Human **Entrez Gene:** 12724 Mouse **Entrez Gene:** 29232 Rat Omim: 600570 Human **SwissProt:** P51788 Human **SwissProt:** Q6IPA9 Human **SwissProt:** Q9R0A1 Mouse **SwissProt:** P35525 Rat **Unigene:** 436847 Human **Unigene:** 177761 Mouse **Unigene:** 11073 Rat

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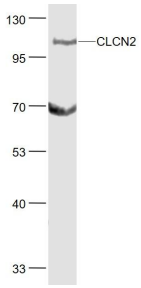
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[VALIDATION IMAGES]



Sample:
HepG2 (Human) Cell Lysate at 30 ug
Primary: Anti- CLCN2 (SL6470R) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 99 kD
Observed band size: 99 kD

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