

Product name:	CLC-7 Rabbit Polyclonal Antibody
Cat number:	ABN08926
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human CLCN7. AA range:10-59
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,ELISA 1:20000-1:40000
Molecular Weight:	90kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

chloride voltage-gated channel 7 (CLCN7) Homo sapiens The product of this gene belongs to the CLC chloride channel family of proteins. Chloride channels play important roles in the plasma membrane and in intracellular organelles. This gene encodes chloride channel 7. Defects in this gene are the cause of osteopetrosis autosomal recessive type 4 (OPTB4), also called infantile malignant osteopetrosis type 2 as well as the cause of autosomal dominant osteopetrosis type 2 (OPTA2), also called autosomal dominant Albers-Schonberg disease or marble disease autosomal dominant. Osteopetrosis is a rare genetic disease characterized by abnormally dense bone, due to defective resorption of immature bone. OPTA2 is the most common form of osteopetrosis, occurring in adolescence or adulthood. [provided by RefSeq, Jul 2008],disease:Defects in CLCN7 are a cause of autosomal dominant osteopetrosis type 2 (OPTA2) [MIM:166600]; also called autosomal dominant Albers-Schonberg disease or marble disease autosomal dominant. OPTA2 is the most common form of osteopetrosis, occurring in adolescence or adulthood. It is characterized by sclerosis, predominantly involving the spine, the pelvis, and the skull base.,disease:Defects in CLCN7 are the cause of osteopetrosis autosomal recessive type 4 (OPTB4) [MIM:611490]; also called infantile malignant osteopetrosis type 2. Osteopetrosis is a rare genetic disease characterized by abnormally dense bone, due to defective resorption of immature bone. The disorder occurs in two forms: a severe autosomal recessive form occurring in utero, infancy, or childhood, and a benign autosomal dominant form occurring in adolescence or adulthood.,function:Mediates the exchange of chloride ions against protons. Functions as antiporter and contributes to the acidification of the lysosome lumen.,miscellaneous:The CLC channel family contains both chloride channels and proton-coupled anion transporters that exchange chloride or another anion for protons. The presence of conserved gating glutamate residues is typical for family members that function as antiporters.,similarity:Belongs to the chloride channel (TC 2.A.49) family.,similarity:Contains 2 CBS domains.,tissue specificity:Brain, testis, muscle and kidney.,