10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

CLCN7 Antibody (C-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP11863B

Product Information

Application	WB, IHC-P, IF, E
Primary Accession	P51798
Other Accession	P51799, O70496, Q4PKH3, NP_001278.1
Reactivity	Human, Mouse
Predicted	Bovine, Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB31756
Calculated MW	88679
Antigen Region	692-720

Additional Information

Gene ID	1186
Other Names	H(+)/Cl(-) exchange transporter 7, Chloride channel 7 alpha subunit, Chloride channel protein 7, ClC-7, CLCN7
Target/Specificity	This CLCN7 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 692-720 amino acids from the C-terminal region of human CLCN7.
Dilution	WB~~1:1000 IHC-P~~1:100~500 IF~~1:10~50 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.05% (V/V) Proclin 300. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	CLCN7 Antibody (C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	CLCN7 (HGNC:2025)
Function	Slowly voltage-gated channel mediating the exchange of chloride ions

against protons (PubMed:18449189, PubMed:21527911). Functions as antiporter and contributes to the acidification of the lysosome lumen and may be involved in maintaining lysosomal pH (PubMed:18449189, PubMed:21527911, PubMed:31155284). The CLC channel family contains both chloride channels and proton-coupled anion transporters that exchange chloride or another anion for protons (By similarity). The presence of conserved gating glutamate residues is typical for family members that function as antiporters (By similarity).

Cellular Location Lysosome membrane; Multi-pass membrane protein

Tissue Location Brain and kidney..

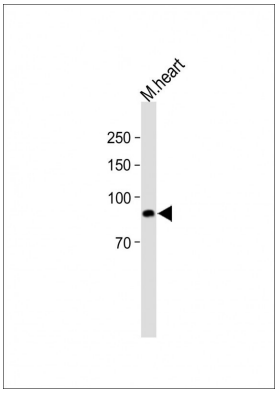
Background

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 autosoml 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.

References

Furthner, D., et al. Klin Padiatr 222(3):180-183(2010)
Phadke, S.R., et al. Indian J. Med. Res. 131, 508-514 (2010) :
Pangrazio, A., et al. Hum. Mutat. 31 (1), E1071-E1080 (2010) :
Kajiya, H., et al. Pflugers Arch. 458(6):1049-1059(2009)
Mazzolari, E., et al. Am. J. Hematol. 84(8):473-479(2009)

Images



All lanes : Anti-CLCN7 Antibody (C-term) at 1:1000 dilution Lane 1: Mouse heart lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated (ASP1615) at 1/15000 dilution. Observed band size : 89kDa Blocking/Dilution buffer: 5% NFDM/TBST.

Citations

- Surface vacuolar ATPase in ameloblastoma contributes to tumor invasion of the jaw bone.