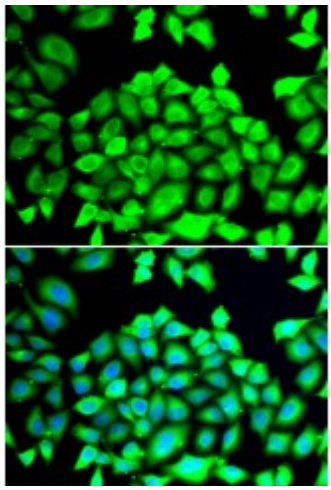
Anti-CLCN7 Antibody



Description	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.

Model	STJ114296
Host	Rabbit
Reactivity	Human
Applications	IF
Immunogen	Recombinant fusion protein containing a sequence corresponding to amino acids 626-805 of human CLCN7 (NP_001278.1).
Gene ID	1186
Gene Symbol	CLCN7
Dilution range	IF 1:50 - 1:100
Tissue Specificity	Brain, testis, muscle and kidney
Purification	Affinity purification
Note	For Research Use Only (RUO).

Protein Name	H(+ /Cl(- exchange transporter 7 Chloride channel 7 alpha subunit Chloride channel protein 7 ClC-7
Molecular Weight	88.679 kDa
Clonality	Polyclonal
Conjugation	Unconjugated
Isotype	IgG
Formulation	PBS with 0.02% sodium azide, 50% glycerol, pH7.3.
Storage Instruction	Store at -20C. Avoid freeze / thaw cycles.
Database Links	HGNC:2025OMIM:166600Reactome:R-HSA-2672351
Alternative Names	H(+ /Cl(- exchange transporter 7 Chloride channel 7 alpha subunit Chloride channel protein 7 ClC-7
Function	Slowly voltage-gated channel mediating the exchange of chloride ions against protons, Functions as antiporter and contributes to the acidification of the lysosome lumen,
Cellular Localization	Lysosome membrane

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