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MFSD8 Rabbit pAb

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Catalog # AP57303

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q8NHS3
Reactivity	Mouse
Predicted	Rat, Pig, Human, Rabbit, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	57628
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MFSD8
Epitope Specificity	301-400/518
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Lysosome membrane.
DISEASE	Defects in MFSD8 are the cause of neuronal ceroid lipofuscinosis type 7 (CLN7) [MIM:610951]. A form of late infantile neuronal ceroid lipofuscinosis. CNL are a clinically and genetically heterogeneous group of neurodegenerative disorders characterized by the intracellular accumulation of autofluorescent lipopigment storage material in different patterns ultrastructurally. The patterns most often observed CLN7 are mixed combinations of granular, curvilinear, fingerprint, and rectilinear profiles. The clinical course includes progressive dementia, seizures, and progressive visual failure.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a ubiquitous integral membrane protein that contains a transporter domain and a major facilitator superfamily (MFS) domain. Other members of the major facilitator superfamily transport small solutes through chemiosmotic ion gradients. The substrate transported by this protein is unknown. The protein likely localizes to lysosomal membranes. Mutations in this gene are correlated with a variant form of late infantile-onset neuronal ceroid lipofuscinoses (vLINCL). [provided by RefSeq, Oct 2008]

Additional Information

Gene ID	256471
Other Names	Major facilitator superfamily domain-containing protein 8, Ceroid-lipofuscinosis neuronal protein 7, MFSD8 {ECO:0000303|PubMed:17564970, ECO:0000312|HGNC:HGNC:28486}

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	MFSD8 {ECO:0000303|PubMed:17564970, ECO:0000312|HGNC:HGNC:28486}
Function	Outward-rectifying chloride channel involved in endolysosomal chloride homeostasis, membrane fusion and function. Conducts chloride currents up to hundreds of picoamperes. Regulates lysosomal calcium content by reducing the lysosomal membrane potential, thereby activating TRPML1 channel and further release of lysosomal calcium ions. Regulates the pH in endolysosomal compartments and may contribute to progressive acidification from endosome to lysosome. Permeable to other halides such as iodide and fluoride ions.
Cellular Location	Endosome membrane; Multi-pass membrane protein. Lysosome membrane; Multi-pass membrane protein. Note=Sorting to lysosomes involves dileucine-based motif
Tissue Location	Expressed at very low levels in all tissues tested.

Background

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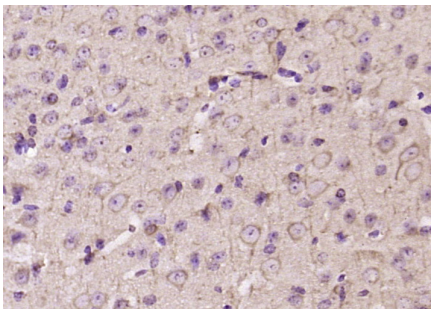
Images



Paraformaldehyde-fixed, paraffin embedded (mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (MFSD8) Polyclonal Antibody, Unconjugated (AP57303) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.