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Rabbit Anti-Junctophilin 3 antibody

SL11083R

Product Name:	Junctophilin 3
Chinese Name:	连接蛋白JPH3抗体
Alias:	JP-3; JP3; JPH3; JPH3_HUMAN; Junctophilin3; Junctophilin type 3; Junctophilin-3; TNRC22; Trinucleotide repeat-containing gene 22 protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Zebrafish,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	81kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Junctophilin 3:181-280/748
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	Junctophilins are components of the junctional complexes between plasma membranes and endoplasmic or sarcoplasmic reticulums present in all excitable cells. Junctophilins contain a cytoplasmic domain which binds to the plasma membrane, as well as an ER/SR membrane spanning hydrophobic C-terminal segment. The three subtypes in this family are Junctophilin-1, -2 and -3. Junctophilin-1 is predominantly expressed in skeletal muscle, but is also expressed at low levels in heart. Junctophilin-2 is expressed

in heart and skeletal muscle. Mutant mice lacking the Jph2 gene exhibit embryonic lethality and possess cardiac myocytes that express abnormal calcium transients. Junctophilin-3 is expressed in brain. The JPH3 alternatively spliced exon 2A has been suggested as a site for CTG repeat expansion leading to a Huntington disease-like autosomal dominant disorder.

Function:
Contributes to the stabilization of the junctional membrane complexes, which are common to excitable cells and mediate cross-talk between cell surface and intracellular ion channels. Probably acts by anchoring the plasma membrane and endoplasmic reticulum. May play an active role in certain neurons involved in motor coordination.

Subcellular Location:
Cell membrane. Endoplasmic reticulum membrane. Localized predominantly on the plasma membrane. The transmembrane domain is anchored in endoplasmic reticulum membrane, while the N-terminal part associates with the plasma membrane.

Tissue Specificity:
Specifically expressed in brain.

DISEASE:
Defects in JPH3 are the cause of Huntington disease-like type 2 (HDL2) [MIM:606438]. Huntington disease (HD) is a neurodegenerative disorder resulting primarily from the loss of medium spiny projection neurons in the striatum, especially in the caudate nucleus, and, to a lesser extent, atrophy of mesencephalic and cortical structures. The typical clinical picture of HD combines familial adult onset chorea and subcortical dementia that usually begin during the fourth decade of life.

Similarity:
Belongs to the junctophilin family. Contains 8 MORN repeats.

SWISS:
Q8WXH2

Gene ID:
57338

Database links:

Entrez Gene: 57338Human

Entrez Gene: 57340Mouse

Entrez Gene: 307916Rat

Omim: 605268Human

	SwissProt: Q8WXH2Human SwissProt: Q9ET77Mouse Unigene: 725123Human Unigene: 306870Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.