

Ankyrin B Rabbit pAb

Catalog # AP98312

Product Information

Application	IHC-P, IHC-F, IF, E
Host	Rabbit
Clonality	Polyclonal
Calculated MW	426 KDa
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Ankyrin B
Epitope Specificity	1901-2100/3957
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 24 ANK repeats.Contains 2 death domains.Contains 2 ZU5 domains.
SUBUNIT	Interacts with RHBG and SPTBN1.
DISEASE	Defects in ANK2 are the cause of long QT syndrome type 4 (LQT4) [MIM:600919]; also known as sick sinus syndrome with bradycardia. Long QT syndromes are heart disorders characterized by a prolonged QT interval on the ECG and polymorphic ventricular arrhythmias. They cause syncope and sudden death in response to exercise or emotional stress. LQT4 displays many atypical features compared to classical long QT syndromes, including pronounced sinus bradycardia, polyphasic T waves and atrial fibrillation. Cardiac repolarization defects may be not as severe as in classical LQT syndromes and prolonged QT interval on EKG is not a consistent feature.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Members of the Ankyrin family of proteins mediate the attachment of integral membrane proteins to the cytoskeleton. ANK1, ANK2 and ANK3 genes encode for the proteins in this family, Ankyrin-1 (also designated Ankyrin R), Ankyrin B and Ankyrin G, respectively. The proteins are structured similarly each composed of an N-terminal domain with multiple ankyrin repeats, a highly conserved central spectrin binding domain, and C-terminal regulatory domains which are susceptible to the most variance. Both Ankyrin B and Ankyrin G are essential for normal neuronal functions. Ankyrin B, or brain Ankyrin is predominantly expressed in the plasma membrane of neurons as well as glial cells throughout the brain. Two transcripts have been described with alternative splicing resulting in additional isoforms. The Ankyrin B protein associates with the spectrin-actin network, mediates axon fasciculation, and stabilizes axon bundles. Ankyrin B is required for coordinated assembly of Na/Ca exchanger, Na/K ATPase, and inositol trisphosphate (InsP(3)) receptor at transverse-tubule/sarcoplasmic reticulum sites in cardiomyocytes.

Additional Information

Gene ID 287

Other Names	ANK-2; CFAP87; FAP87; LQT4; brank-2; ankyrin 2; long (electrocardiographic) QT syndrome 4; ankyrin 2, neuronal
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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.
Name	Ankyrin B Rabbit pAb
Function	Attaches integral membrane proteins to cytoskeletal elements. Also binds to cytoskeletal proteins. Required for coordinate assembly of Na/Ca exchanger, Na/K ATPase and InsP3 receptor at sarcoplasmic reticulum sites in cardiomyocytes. Required for the coordinated expression of the Na/K ATPase, Na/Ca exchanger and beta-2-spectrin (SPTBN1) in the inner segment of rod photoreceptors. Required for expression and targeting of SPTBN1 in neonatal cardiomyocytes and for the regulation of neonatal cardiomyocyte contraction rate.

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